

Sickle Cell Disease And Thalassaemia

Information on pre-travel preparation, tips to stay health abroad and links to useful resources for travellers with sickle cell disease and thalassaemia

Key Messages

Travellers with sickle cell disease and some thalassaemias are at increased risk of infection.

Severe malaria is also a risk for these travellers. If travel is unavoidable, bite avoidance and strict adherence to anti-malarial prophylaxis is essential.

Any fever should be urgently evaluated due to risk of bacterial sepsis or malaria.

Travellers with sickle cell disease need to be aware of triggers for sickle cell crisis and know how to minimise the risks.

The risk of sickle cell crisis increases at altitude, when oxygen concentration in air is reduced. Travel to altitudes above 2,000 metres may trigger a crisis. During air travel, supplemental oxygen may be recommended for some.

Travellers carrying opioid pain medication may need to declare this to the airline and consult the destination country to ensure there are no entry restrictions.

Overview

Sickle cell disease (SCD) and thalassaemia are inherited red blood cell disorders, caused by the presence of an abnormal form of haemoglobin (oxygen transporting protein). According to the World Health Organization (WHO), approximately 5% of the world's population are healthy carriers of either of these genes [1].

Sickle cell disease and sickle cell trait

Individuals with SCD have red blood cells that are 'sickle' shaped (like a crescent moon) instead of the normal 'doughnut' shape. SCD presents when the gene for sickle haemoglobin (HbS) is inherited from both parents. This is termed homozygous sickle cell disease (HbSS) or sickle cell anaemia [2]. This is the most common and severe type of SCD. Those who inherit a sickle cell gene together with a gene for normal haemoglobin (HbA), have sickle cell trait (also known as being a

sickle cell 'carrier'). It is rare for sickle cell trait to cause health problems [2].

Sickle cell disease mainly affects those whose family origins are African or African-Caribbean.[2].

When oxygen is released into tissues, sickle cells change shape and stick together. Because of their shape they are unable to pass through small blood vessels, blocking the vessel and causing pain, usually in bones and joints. This can happen suddenly and be severe, lasting up to a week, causing symptoms known as sickle cell crisis. Triggers include the cold, lack of oxygen, dehydration, hard exercise, fever and infection [2]. Repeated blockages of blood vessels in, for example, the liver, kidney and spleen, can result in these organs being damaged. Those with SCD, particularly children, are also at increased risk of serious infections, mainly due to either partial failure of the spleen (hyposplenism) or complete failure (asplenia) [2].

Sickle cells are also removed from the body more easily than healthy red blood cells, often resulting in anaemia. Blood transfusions can decrease anaemia and reduce sickling temporarily [2, 3].

Treatment for sickle cell conditions involves the prevention and management of both infection and complications resulting from blockage of blood vessels. Stem cell transplant is the only available treatment that can cure SCD, and is only used in severe cases [2]. To reduce the number of painful crises in those with SCD, hydroxycarbamide (hydroxyurea) may be recommended [2, 4]. However, hydroxycarbamide can have side effects and needs specialist monitoring [2].

Thalassaemia (alpha and beta) and thalassaemia trait

Thalassaemias are inherited conditions resulting in a reduced production of normal haemoglobin and the breakdown of red blood cells, leading to anaemia [5]. As a result, the body tries to produce more haemoglobin and red blood cells, which can cause additional complications [5].

Thalassaemias are classified as alpha and beta thalassaemias, according to which part (alpha or beta) of the haemoglobin is affected. Both alpha and beta thalassaemia are classified further according to severity of the condition e.g. major, intermediate or minor.

Beta thalassaemia major is the most severe type and causes severe anaemia, requiring regular blood transfusions and iron chelation therapy (removal of excess iron). In addition to anaemia, complications include iron overload (due to repeated blood transfusions), heart arrhythmias, hormone complications (leading to delayed puberty and restricted growth), liver disease and death [5]. The increased red blood cell production can result in enlargement of the spleen and reduced ability to control infections if challenged.

Treatment may be necessary for some beta thalassaemias, depending on severity of disease [5]. Stem cell or bone marrow transplant is currently the curative treatment option but is rarely performed [5].

Those with thalassaemia trait, who carry only one abnormal thalassaemia gene rather than two,

may not have any symptoms or mild anaemia only.

Beta thalassaemia mainly affects people of Mediterranean, South Asian, Southeast Asian and Middle Eastern origin. Alpha thalassaemia is common in South East Asia, Africa and India [5].

COVID-19

Current advice is that anyone with a weakened immune system is clinically vulnerable to severe COVID-19. Those with rare diseases that significantly increase the risk of infections, including homozygous sickle cell disease, are considered clinically extremely vulnerable [6]. Further specific advice for those with haemoglobin disorders is provided by the [British Society for Haematology](#) and should be read alongside [current government guidance](#).

All individuals, but particularly those who are clinically vulnerable or clinically extremely vulnerable to severe COVID-19 disease, need to consider [current UK recommendations](#) to reduce their risk of infection, which may mean postponing travel.

General guidance regarding [risk assessment](#) for travel during the COVID-19 pandemic and information about the [COVID-19 vaccination programme](#) is available.

Pre-travel preparation

Advice for those with SCD or thalassaemia should follow the usual advice given to all travellers, and tailored to the individual, considering severity of their illness, current treatment and medications. Assessment of a travellers' risk and the suitability of their destination and plans should ideally be discussed with a healthcare professional (and/or specialist) prior to booking. SCD and thalassaemia may be associated with reduced function of the spleen, and will need to be assessed on an individual basis. For those with a dysfunctional or absent spleen, advice on the need for additional immunisations and antibiotic prophylaxis and standby treatment is [available](#).

Destination specific risks and how to reduce risks can be found on the [Country Information pages](#). Malaria is considered a significant cause of illness and death in those with SCD. Those with no spleen or severely impaired splenic function are also at particular risk of severe malaria. Where possible, travel to a malarious areas should be avoided [7].

Although little data is available, studies suggest that those with SCD can be at greater risk of medical complications when abroad [3]. Infection has been documented as the main hazard for children with SCD when travelling, primarily due to reduced function of the spleen [8]. However, both SCD and some thalassaemias are associated with an increased risk of infection for a variety of reasons, including splenic dysfunction; but also due to iron overload in those with thalassaemia [5]. Travellers should therefore ensure that any fever is urgently evaluated due to increased risk of bacterial sepsis and complications of malaria if infected.

In addition to the increased risk of infection, the main risk for adult travellers with SCD appears to

be sickle cell crises (blockage of blood vessels) [3]. Travellers with SCD should be aware of the triggers for crises, and know how to minimise the risks. They should also know where to access urgent medical care whilst abroad in the event of a sickle cell crisis or other complications. The [Global Sickle Cell Disease Network](#) lists researchers and clinicians worldwide working together to assist those with SCD.

Travellers should be aware that blood transfusions may not be reliable, nor available in many countries. Travel plans may need to be co-ordinated with transfusion schedules and iron chelation therapy in those with thalassaemia needing regular treatments.

Carrying a letter from a haematologist with the diagnosis and current drug treatment may be beneficial; this is particularly important if carrying opioid pain medications, which may need to be declared to the airline. Travellers should also determine if there are any restrictions on medications that can be carried into their destination country. Further information is available in the [Medicines and Travel](#) factsheet.

Travellers should also be advised to carry their own necessary medication, as substandard or falsified medication can be a problem in some countries.

Comprehensive travel insurance is essential for all travellers. A full declaration of medical conditions should be made to the insurers and all equipment and planned activities should also be covered.

Journey risks

Flying

Sickle cell disease may be exacerbated by reduced oxygen pressures present in the air cabins. Because a crisis could be life-threatening, supplemental oxygen is sometimes recommended in-flight for those with SCD [9, 10]. This would need to be arranged with the airline prior to travel.

Sickle cell trait does not usually present a particular problem at normal cruising altitude in a pressurised aircraft, although there are rare reports of splenic infarction following air travel [11].

Travel should be deferred for at least 10 days following a sickle crisis and may need assessment by a clinician with aviation medicine experience and a medical assessment of fitness to fly [10, 12].

Severe anaemia should be corrected prior to air travel, and for those with a haemoglobin level less than 7.5g/dl, a medical assessment of fitness to fly would be needed, and oxygen considered in-flight [10].

Venous Thromboembolism

Prolonged air travel is a risk for venous thromboembolism (VTE) and studies from the WHO

Research into Global Hazards of Travel (WRIGHT) project indicates that the risk of VTE approximately doubles after a long-haul flight (> 4 hours), and also with other forms of travel where travellers are seated and immobile [13]. Those with SCD are at increased risk of venous thromboembolic disease, therefore an individual assessment of risk factors for thrombosis is required for these travellers.

Travellers should therefore be advised on measures to prevent travel related VTE including maintaining mobility in-flight [14]. Additional information on reducing the risk of VTE can be found on the [Venous Thromboembolism](#) factsheet.

Food and water borne risks

Travellers with SCD are more susceptible to developing hypovolaemia (a reduced volume of blood circulating in the body) because of disease of small blood vessels in the kidney [3, 15]. Hypovolaemia may be further exacerbated by sun or heat exposure, lack of access to safe drinking water and diarrhoeal disease. This in turn could lead to sickle cell crisis during travel. It is therefore essential to advise on making safe food choices during travel and to maintain adequate hydration.

Due to an increased risk of infection, those with SCD and some thalassaemias should also have a lower threshold for seeking medical attention for diarrhoeal illness [3].

After careful risk assessment, standby antibiotics to be used in the event of diarrhoea may be considered when travelling to high-risk countries [16, 17]. Further information can be found in the [Travellers' diarrhoea](#) factsheet.

Vector borne risks

Babesiosis, a parasitic infection transmitted by ticks, can result in severe infection in those with an absent or dysfunctional spleen. Bacterial infection with *Capnocytophaga* and *Pasturella* (both associated with cat and dog bites) can also cause overwhelming infection in some of these travellers. Further information on specific vector-borne risks for travellers with SCD or thalassaemias and associated splenic dysfunction can be found in the [Asplenia and Hyposplenia](#) factsheet.

Malaria

[Malaria](#) is considered a significant cause of illness and death in those with sickle cell disease, causing further destruction of red blood cells in addition to that due to sickle cell disease itself [7, 18]. It is therefore essential that travellers with sickle cell disease who visit countries / areas with a risk of malaria take rigorous mosquito bite avoidance measures.

Sickle cell trait confers some protection against malaria, although antimalarial prophylaxis would still be required for those visiting areas at risk for malaria [7].

Thalassaemia may provide protection against severe malaria, but there is no evidence that it prevents uncomplicated malaria [7].

Splenic function must also be considered for those with SCD and thalassaemia. For those without a spleen or if splenic function is severely impaired, then rigorous use of mosquito bite prevention measures and strict adherence to appropriate chemoprophylaxis (malaria tablets) is advised, even in low risk and very low risk malaria areas, where bite avoidance and awareness alone are recommended for other travellers [7].

Vaccination

Those with SCD and thalassaemias should be up to date with routine immunisations and boosters as [recommended in the UK](#). In addition, immunisations for those with splenic dysfunction need to be considered (i.e. pneumococcal, Men ACWY, Men B and influenza) [19]. Those receiving regular blood transfusions should be vaccinated against hepatitis B [20].

Sickle cell disease and thalassaemia alone are not considered a contraindication for vaccination [3]. However, as in all travel consultations it is important to consider any other underlying medical conditions, treatment or drug therapy which may contraindicate the use of live vaccines. Health professionals are encouraged to seek expert advice if further information is needed regarding use of live vaccines.

Other health risks

Altitude

For those with SCD, slight reduction in oxygen concentration at altitude can induce sickling. There is an increased risk of sickle cell crisis or splenic infarcts at altitudes above 2,000 metres. Travel to altitude should therefore be avoided in those with SCD [21].

Travellers with sickle cell trait and some thalassaemias (e.g. beta thalassaemia), are also at risk of crisis due to low oxygen concentration at high altitude. Splenic infarct has been noted at high altitudes in those with sickle cell trait, and severe exertion has also been associated with sickle cell crisis and sudden death in this group of individuals [21, 22]. Those with sickle cell trait who decide to travel to altitude should therefore be aware of the risks and advised to avoid over-exertion, maintain adequate hydration and to minimise heat stress [23, 24].

General advice for those who get sick abroad

Travellers should seek medical attention urgently if they have signs of infection (particularly fever), have had animal contact such as a bite, scratch or lick to an open wound, or a tick bite in a country with risk of tick-borne disease.

When seeking medical care abroad, travellers should inform health professionals if they have a dysfunctional spleen, together with other background medical information.

Resources

- [NHS. Sickle Cell Disease](#)
- [Global Sickle Cell Disease Network](#)
- [NHS. Thalassaemia](#)
- [Thalassaemia International Federation](#)
- [British Society for Haematology](#)
- [GOV.UK: Coronavirus \(COVID-19\)](#)

Patient Associations

- [Sickle Cell Society](#)
- [UK Thalassaemia Society](#)

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