The Royal College of Pathologists
Pathology: the science behind the cure

National Medical Examiner's Good Practice Series No. 20

Sickle cell disease

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About the National Medical Examiner's Good Practice Series

Medical examiners are senior doctors providing independent scrutiny of non-coronial deaths in England and Wales, with the role now a statutory requirement since 9 September 2024.

While there is extensive guidance available on a wide range of topics for NHS and public sector staff, the National Medical Examiner's Good Practice Series highlights how medical examiners and medical examiner officers can better meet the needs of local communities and work more effectively with colleagues and partners.

The Good Practice Series is a topical collection of focused summary documents, designed to be easily read and digested by busy front-line staff, with links to further reading, guidance and support.

Introduction

Medical examiners cannot be expert in every disease or condition that causes or contributes to death, but some diseases are noteworthy as they are consistently linked to inequalities in treatment that can lead to premature death. Sickle cell disease is one example and the level of awareness of these issues varies among non-specialists.[1]

There is a national approach to the registration, diagnosis and management of sickle cell disease. The National Haemoglobinopathy Panel[2] works to improve services, equity and patient experience by working in collaboration with Haemoglobinopathies Coordinating Centres and other stakeholders in England. The National Haemoglobinopathy Registry[3] is a database of patients with red cell disorders. National Institute for Health and Care Excellence (NICE) also provides guidance for reasonable standards of care,[4] including providing analgesia for all people presenting with an acute, painful sickle cell episode within 30 minutes of presentation in secondary care.

Some patient and public information is reproduced in this document and may improve awareness of sickle cell disease among medical examiners and medical examiner officers. This will assist medical examiners in establishing accurate causes of death and consistent coroner notifications, and identifying opportunities for learning and improvement.

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[1] NHS. *Sickle cell and thalassaemia. List of high and low prevalence Trusts: England.* Available at: https://assets.publishing.service.gov.uk/media/5a7f4a17ed915d74e33f583a/HP_and_LP_Trusts_2015.pdf
[2] NHS. *National Haemoglobinopathy Panel*. Available at: https://www.nationalhaempanel-nhs.net/
[3] NHR. *National Haemoglobinopathy Register*. Available at: https://nhr.mdsas.com/
[4] NICE. *Sickle cell disease*. Available at: https://cks.nice.org.uk/topics/sickle-cell-disease/

Recommendations for medical examiners

Medical examiners and officers should:

1. consider that sickle cell disease is likely have contributed to the causes of death. The opinion of an expert haematologist (who may be employed at tertiary care centres) is likely to be helpful to determine whether or the extent to which this was the case
2. be alert for opportunities for learning and improvement in care for patients as clinicians' lack of understanding and training in relation to sickle cell disease has been highlighted in case record reviews and coroners' reports
3. examine areas such as recognition of sickle cell crises, timeliness of interventions such as analgesia and transfusion and, where treatment took place at smaller haematology centres without sickle cell facilities, whether referral to specialist centres was considered appropriately or whether there was adequate communication with a haematologist
4. pay close attention to concerns highlighted by families and carers as they will be familiar with the patient, how they managed sickle cell disease and their experience of care
5. note that in some communities, the cultural stigma attached to sickle cell disease may make relatives less likely to raise or pursue concerns that could contribute to improving care for future patients
6. where it appears omissions in care may have occurred, or where the patient may not have followed advice about managing their condition, consider carefully whether notification to the coroner is required under the Notification of Deaths Regulations 2019 and ensure the coroner is provided with all the available information
7. if the coroner decides not to investigate the death, consider the interests of the bereaved carefully and liaise closely with the coroner's office to minimise delays. If the attending practitioner or medical examiner is unable to certify the death to the best of their knowledge and belief with the information available to them, notification to the coroner is appropriate for proper establishment of the causes of death.

Context and background

The Sickle Cell Society provides a range of helpful information, which is summarised in this section, along with information from NHS A–Z.

People with sickle cell disease are born with the condition, which is a disorder of the haemoglobin in the red blood cells. It can only be inherited from both parents, each having passed on the gene for sickle cell, and is not contagious. It is more common in people with an African or Caribbean family background.

Symptoms of sickle cell disease include anaemia, an increased risk of serious infections and episodes of severe pain known as crises. The pain occurs when the cells change shape after oxygen has been released. The red blood cells then stick together, causing blockages in the small blood vessels. These painful episodes are treated with strong painkillers, such as morphine. The Royal College of Emergency Medicine has guidance for managing acute presentation of sickle cell crises.

People with sickle cell disease are at risk of complications including stroke, acute chest syndrome, blindness, bone damage and priapism. Over time, people with sickle cell disease can experience damage to organs such as the liver, kidney, lungs, heart and spleen. Death can also result from complications of the disease. Treatment of sickle cell disease mostly focuses on preventing and treating complications.

For many years, the only cure for sickle cell disease has been bone marrow/stem cell transplant, but this is only possible for a limited number of affected individuals who have a suitable donor (usually a full sibling). A new gene therapy treatment (Casgevy, or Exa-Cel[5]) has recently been approved by NICE but will only be available to approximately 50–60 patients a year.

People with sickle cell disease need treatment throughout their lives. This is usually delivered by different health professionals in a specialist sickle cell treatment centre. It is also important for people with sickle cell disease to look after their own health using self-

[5] NICE. *Exagamglogene autotemcel for treating severe sickle cell disease in people 12 years and over*. Available at: https://www.nice.org.uk/guidance/ta1044/chapter/2-Information-about-exagamglogene-autotemcel

care measures, such as by avoiding triggers and managing pain. A number of interventions to manage sickle cell disease are available, including:

- drinking plenty of fluids and staying warm to prevent painful episodes
- painkillers, such as paracetamol or ibuprofen (sometimes treatment with stronger painkillers in hospital may be necessary; NICE guidance is provision of analgesia within 30 minutes for all people presenting with an acute painful sickle cell episode in secondary care)
- daily antibiotics and having regular vaccinations to reduce chances of an infection
- hydroxycarbamide (hydroxyurea) to reduce symptoms
- regular blood transfusions if symptoms continue or get worse, or there are signs of damage caused by sickle cell disease
- an emergency blood transfusion if severe anaemia develops.

NICE noted in a UK study that the median survival is estimated at 67 years in people with sickle cell anaemia and higher in people with the Hb SC genotype, while a US study reported a median survival of 58 years for people with sickle cell anaemia and 66 years for those with Hb SC disease.

The NHS A–Z notes that the life expectancy for someone with sickle cell disease tends to be shorter than normal, but this can vary depending on the exact type of sickle cell disease they have, how it is treated and what problems they experience.

A study in The Lancet revealed a high contribution of sickle cell disease to all-cause mortality that is not apparent when each death is assigned to only a single cause. It noted that, as a cause of death, sickle cell disease is often underreported and underdiagnosed. Medical examiners need to be aware of the increased risk of infection and of death from complications like stroke, cardiac and renal disease, and pregnancy morbidity.

Report from the All-Party Parliamentary Group on Sickle Cell and Thalassaemia and the Sickle Cell Society

The No-one's Listening report was triggered by the coroner's report into the death of Evan Nathan Smith. Key findings include:

- evidence of sub-standard care for sickle cell disease patients admitted to general wards or attending Accident & Emergency (A&E) departments (including lack of adherence to national care standards)
- low awareness of sickle cell disease among healthcare professionals and examples of inadequate training and insufficient investment in sickle cell disease care
- reports of negative attitudes towards sickle cell patients and evidence suggesting such attitudes are often underpinned by racism.

These findings align to the Health Services Safety Investigations Body (HSSIB) investigation report[6] concerning the management of SCD. It is significant that such shortcomings can occur in NHS providers serving communities with demographic profiles and ethnic populations where the disease is more common. If oversights are possible for such providers, they are even more likely where the population served means NHS staff are likely to encounter fewer patients with sickle cell disease and to be less aware of the disorder.

Considerations for medical examiners

It is very likely that sickle cell disease will have contributed to deaths of those with the disorder. It is appropriate for medical examiners to actively consider this possibility and to question events and care in the relevant period before the death. Given the specialist nature of care for those with the disorder, medical examiners will often benefit from insights from consultant haematologists and other clinicians with expert understanding of care for patients with sickle cell disease. This may require liaising with colleagues beyond those who were caring directly for the deceased.

Families and carers will be able to provide valuable insights, as often they become familiar at close range with how the deceased managed their health, and with the care and support provided by the NHS. Concerns and issues identified in discussion with the bereaved will be of great value in determining whether a death should be referred for clinical governance review or notified to the coroner.

[6] HSSIB. *Management of sickle cell crisis.* Available at: https://www.hssib.org.uk/patient-safety-investigations/management-of-sickle-cell-crisis/

However, it should also be noted that, in some communities, having sickle cell disease or carrying sickle cell trait can bring social stigma. In some cases, there may a desire to avoid drawing attention to sickle cell disease playing a part in a death, for example as part of a clinical governance review or coroner notification. There will be a need for sensitive discussion and communication, though the requirement for medical practitioners (including medical examiner) to notify certain deaths to the coroner is statutory and cannot be applied differently when it applies.

Find out more

- NHS – NHS trusts with high and low prevalence of sickle cell disease.
- HSSIB – Management of sickle-cell crisis.
- National Haemoglobinopathy Panel.
- National Haemoglobinopathy Register.
- National Confidential Enquiry into Patient Outcome and Death report – Sickle: A sickle crisis? (2008).
- NICE – Sickle cell disease.
- Royal College of Emergency Medicine – Management of presentations of acute sickle cell disease in the emergency department.
- Royal College of Pathologists:
  - Guidelines on autopsy practice: Autopsy in sickle cell disease and sickle trait
  - Autopsy webinar series – Sickle cell disease.
- Sickle Cell Society.

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