



SPAHA

SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Data Quick Guide

Haemoglobinopathy Database (CAS)

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July 2021, Information Management Service

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Searching Patients

Using CHI number

The quickest way to search for a patient is to search for them by their CHI number.

Patient	Reports	CHI : <input type="text"/>	Search
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Alternatively patients can be found using **two** pieces of information, such as forename and gender.

Surname	<input type="text"/>	Gender	Male <input type="button" value="v"/>
Forename	Thomas	Postcode	<input type="text"/>
Date of Birth	<input type="text"/>	Hospital Number	<input type="text"/>

Find

Note: When searching with two pieces of information, only patients from **your** base treatment centre will be returned, unless all centres are selected.

CHI	Gender	Forename	Surname	Address	Treatment Centre
111111111	Male	THOMAS	RUTHERFORD	24 BREAD STREET EDINBURGH	Edinburgh - Children

Click on the patient in the list and press the  View button to open the patient record.

Adding Patients

Importing from SCI store

Patient
Find Patient
Add Patient

To add a patient to the database, use the menu to select the **Add Patient** option.

Enter the patients CHI number into the 'CHI' field and press [Import](#) to import the demographic information from SCI store (the centralised place which links to GP information systems).

THOMAS RUTHERFORD

CHI: 1111111111
Gender: M
Date Of Birth: 11/11/1911
Address: 24 BREAD STREET EDINBURGH
Postcode: EH3 9AF

Add

Press the **add** button to copy the details into the database. Finally, enter the patient's health board of residence.

Consent

As consent is no longer required there is no need to complete this box. This will be removed altogether from CAS in due course

Patient Consent

Consent given? ?

Date of consent ?

Indicator 1

Seen within 8 weeks

100% of screen positive babies are seen by a paediatric haematologist or paediatrician within 8 weeks of referral from Newborn Screening

Background

Early communication with the local named healthcare professional is important to provide accurate information and to ensure that the infant has timely access to prophylactic treatment.

Sickle cell disease in childhood: standards and guidelines for clinical care published by the NHS Sickle Cell and Thalassaemia Screening Programme in partnership with the Sickle Cell Society [PHE, 2010].



Data Entry Steps

The date when the family is seen is recorded under the **core dataset** tab (see below):

The screenshot shows a navigation bar with tabs: Demographics, Conditions, Encounters, Results, Medications, and Core Dataset. The 'Core Dataset' tab is selected. Below it is a form with three sub-tabs: General, Confirmatory Results, and Other. The 'General' sub-tab is active. The form contains the following fields:

- Date of blood spot specimen
- Date blood spot tested
- Result (dropdown menu: Select an Option)
- Date reported to clinician (text input field, circled in red)
- Haematologist / Paediatrician (dropdown menu: Select an Option)
- Follow up form received (radio buttons: Yes, No)
- Date parents contacted
- Date parents seen (text input field, circled in red)

A red dashed box highlights the 'Date reported to clinician' and 'Date parents seen' fields, with arrows pointing to a callout box containing the text: 'Date reported to clinician' and 'Date parents seen'. Below the form is a 'Save' button.

Note: please ensure you press save before moving on to other tabs to ensure data is not lost.

Patients born in this reporting year can be found in the quarterly data update report for your centre.

Indicator 2

NBS Lab Informed

100% of screen positive babies in whom results of confirmatory testing are returned to the New Born Screening Laboratory and New Born Screening will update CAS as such.

Background

Ensure that screening is backed up by a diagnostic test.

Note: This Data will be completed by the New Born Screening



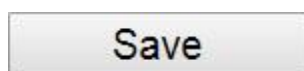
Data Entry Steps

The results of confirmatory testing being returned to the NBS lab are recorded under the **core dataset** tab (see below).



From the '**General**' tab locate the appropriate field.

Note: please ensure you press save before moving on to other tabs to ensure data is not lost.



Patients born in this reporting year can be found in the quarterly data update report for your centre

Indicator 3

Antibiotic prophylaxis

100% of infants with Sickle Cell disease are offered penicillin V (or alternative) by 3 months of age

Background

People with sickle cell disease are particularly susceptible to infections, especially of the respiratory tract and septicaemia. Lifelong antibiotic prophylaxis is therefore recommended for all people with sickle cell disease and it is particularly important that there is full adherence up to 5 yrs of age.


Sickle cell disease in childhood: standards and guidelines for clinical care published by the NHS Sickle Cell and Thalassaemia Screening Programme in partnership with the Sickle Cell Society (PHE, 2010)

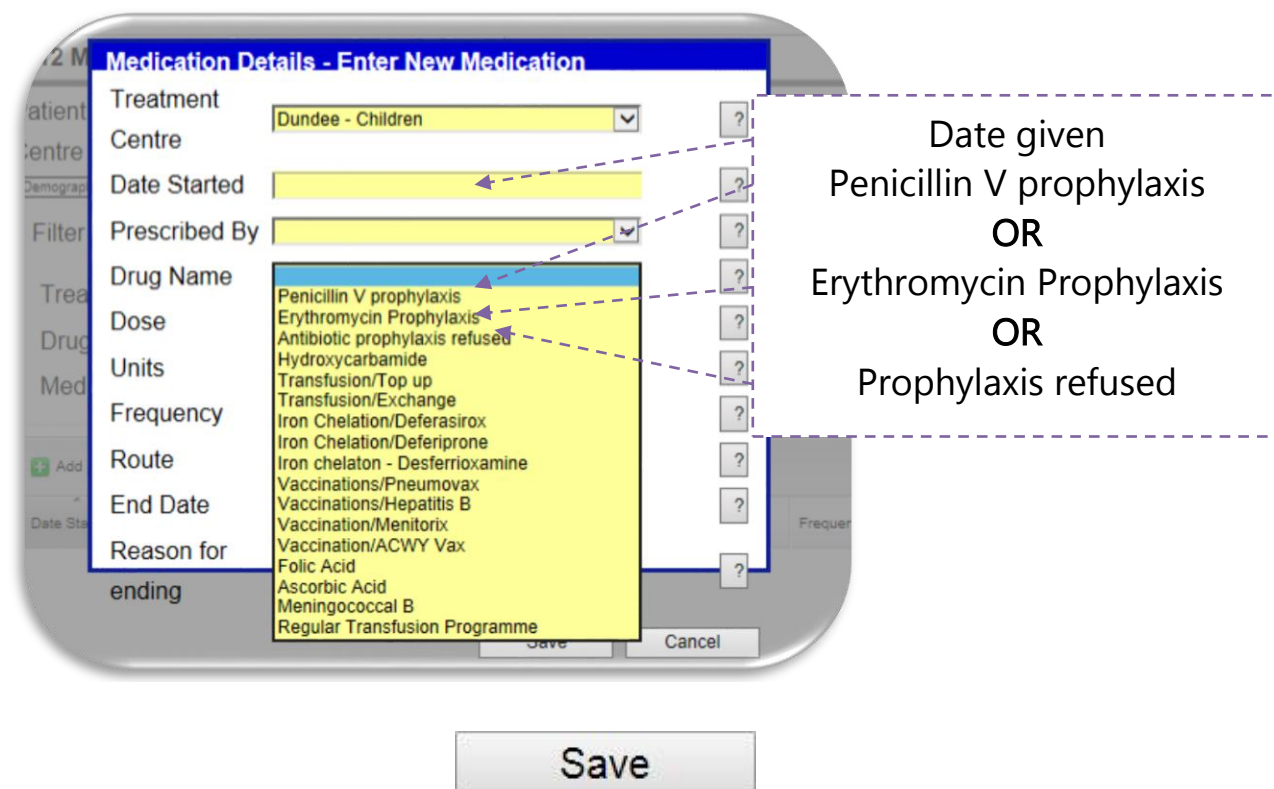


Data Entry Steps

Antibiotic prophylaxis is recorded under the **medications** tab (see below)



To add a new medication, click the  **Add** button.



Medication Details - Enter New Medication

Treatment: Dundee - Children

Centre: Dundee - Children

Date Started: [Date field]

Prescribed By: [User dropdown]

Drug Name: Penicillin V prophylaxis, Erythromycin Prophylaxis, Antibiotic prophylaxis refused, Hydroxycarbamide, Transfusion/Top up, Transfusion/Exchange, Iron Chelation/Deferasirox, Iron Chelation/Deferiprone, Iron chelaton - Desferrioxamine, Vaccinations/Pneumovax, Vaccinations/Hepatitis B, Vaccination/Menitorix, Vaccination/ACWY Vax, Folic Acid, Ascorbic Acid, Meningococcal B, Regular Transfusion Programme

Date given
Penicillin V prophylaxis
OR
Erythromycin Prophylaxis
OR
Prophylaxis refused

Save

Note: please ensure you press save before moving on to other tabs to ensure data is not lost.

Patients turning 3 months old in this reporting year can be found in the quarterly data update report for your centre

Indicator 4

Pneumovax

95% of patients should be offered Pneumovax (polysaccharide antigen) at 2 years of age and 5 yearly thereafter

Background


The immunization recommendations are largely based on expert opinion in the guidelines *Sickle cell disease in childhood: standards and guidelines for clinical care* published by the NHS Sickle Cell and Thalassaemia Screening Programme in partnership with the Sickle Cell Society [PHE, 2010] and *Standards for the clinical care of adults with sickle cell disease in the UK published by the Sickle Cell Society* [Sickle Cell Society, 2008], and in a British Medical Journal (BMJ) review article on sickle cell anaemia [Brousse et al., 2014].

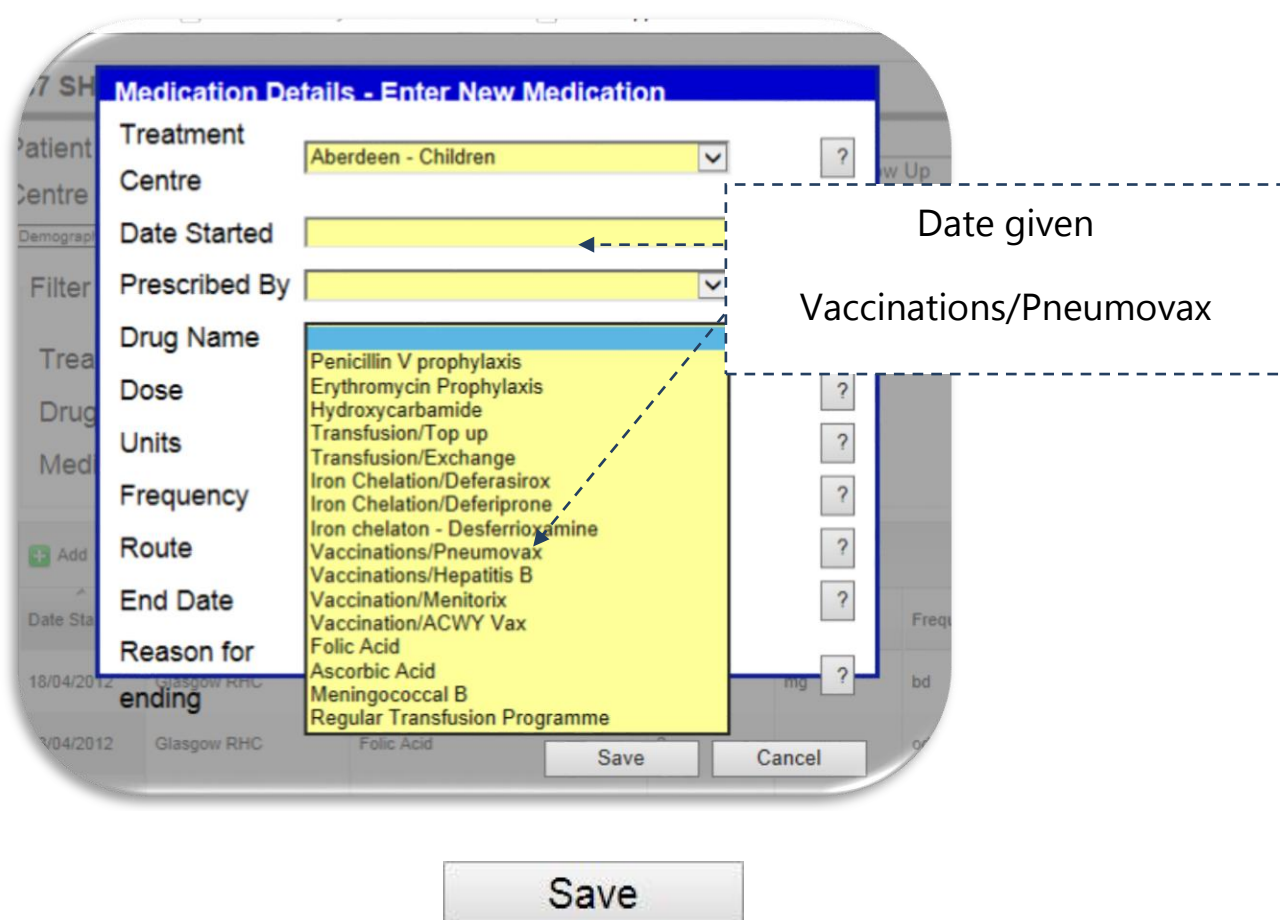


Data Entry Steps

Pneumovax is recorded under the **medications** tab (see below).



To add a new medication, click the  **Add** button.



Medication Details - Enter New Medication

Treatment: Aberdeen - Children

Centre: [Dropdown]

Date Started: [Date Picker]

Prescribed By: [Dropdown]

Drug Name: Penicillin V prophylaxis, Erythromycin Prophylaxis, Hydroxycarbamide, Transfusion/Top up, Transfusion/Exchange, Iron Chelation/Deferasirox, Iron Chelation/Deferiprone, Iron chelaton - Desferrioxamine, **Vaccinations/Pneumovax**, Vaccinations/Hepatitis B, Vaccination/Menitorix, Vaccination/ACWY Vax, Folic Acid, Ascorbic Acid, Meningococcal B, Regular Transfusion Programme

Dose: [Dropdown]

Units: [Dropdown]

Frequency: [Dropdown]

Route: [Dropdown]

End Date: [Date Picker]

Reason for ending: [Dropdown]

Buttons: Save, Cancel

Annotations: Date given, Vaccinations/Pneumovax

Bottom Button: Save

Note: please ensure you press save before moving on to ensure data is not lost.

Patients due first or booster doses this reporting year can be found in the quarterly data update report for your centre. Data for patients who have received their vaccination at clinic should be entered and efforts made to retrieve information for patients whose vaccination may have been delivered by a GP.

Indicator 5

Transcranial Doppler Scanning (TCD)

100% of children with Haemoglobin SS aged 2-16 years offered an annual TCD scan

Background

Annual TCD scans should be performed on all children with SCD from 2 years in accordance with the TCD Standards and Guidelines. For those children who are considered to be 'high risk', the risks and benefits of starting regular blood transfusions and/or other treatments should be fully discussed with parents/carers.

Note: This information is usually entered by the TCD service in Glasgow, however if your patient has declined the offer this should be entered locally.



Data Entry Steps

Demographics Conditions Encounters **Results** Medications Core Dataset



1. Results / Radiology / TCD

Result Details - Enter New Result	
Treatment Centre	Edinburgh - Children
Requested By Clinician	
Date of Investigation	
Discipline	Radiology
Investigation	
Result	MRI - Cardiac TCD MRI - Liver Ferriscan (Paediatric) Ferriscan (Adult) TCD Offered

Result Summary - Free text area mentioning:

- "Normal/Abnormal/Conditional"
- "Asymmetric/Insufficient/Uncooperative"

And the velocities, "LMCA 123 RMCA 132"

Result Action - Free text area mentioning year/months

- "Repeat 2 months"
- "Repeat 1 year"

Save

Demographics Conditions **Encounters** Results Medications Core Dataset

2. Outpatient Care, Clinical Review, Encounter Summary free text area

Indicator 6

Iron overload monitoring

90% of thalassemia patients or sickle cell patients on regular long-term transfusion (1 year and over) undergoing appropriate monitoring of iron overload (annual MRI as per guidelines).

Background

Achieving optimal outcomes depends on effective use of monitoring tests to modify chelation regimes to target both liver and cardiac iron. MRI is used to monitor iron burden in both the liver and the heart.

Treatment of iron overload for transfused and non-transfused patients with chronic inherited anaemias. [NHS England: 16070/P].

UK Thalassemia Society – Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK.

Management of iron overload in children. BJH 2014.

NOTE: Two data items have to be recorded for this audit.

1. Patient receives regular transfusion
2. Date of MRIs for these patients



A regular transfusion programme is recorded using the **medications** tab:



Regular transfusion programme (indicates date when regular transfusion started)

Ferriscan or MRI data is recorded in the **results** tab under radiology:



MRI to monitor iron overload:
MRI Cardiac
MRI Liver
Ferriscan (Paediatric)
Ferriscan (Adult)

Indicator 7

Annual Review

100% of patients with Thalassaemia or Sickle Cell disease should be offered an annual review.

Background

Patients with Thalassaemia require input from a range of specialist health professionals. According to *Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK*, patients with Thalassaemia should be reviewed by an appropriate specialist at least once a year to offer access to optimal specialist care regardless of where they live.

In *Sickle Cell disease in childhood: Standards and Guidelines for Clinical Care*, one role of the specialist healthcare team is to carry out an annual review of all children with Sickle Cell Disease.

Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK, 3rd Edition.


Sickle Cell disease in childhood: Standards and Guidelines for Clinical Care. 2nd edition October 2010.



Data Entry Steps

Demographics Conditions **Encounters** Results Medications Core Dataset



To add a new encounter, click the  **Add** button.

Encounter Details - Enter New Encounter	
Treatment Centre	Glasgow RHC
Start Date	
Patient Attendance	

Enter the date of the annual review as the '**Start date**'.

Annual review can be selected by choosing '**Outpatient Care**' and then '**Annual review**'.

Encounter Type	Outpatient Care ▼
Encounter Reason	Annual Review ▼

Save

Note: please ensure you press save before moving on to ensure data is not lost.

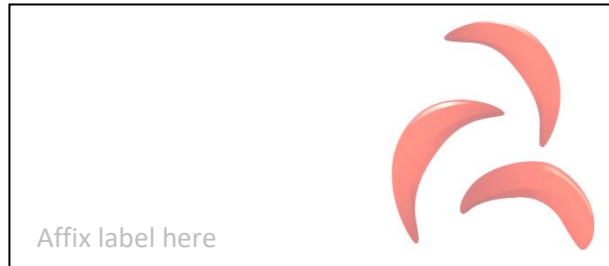
Patients due for an annual review in this reporting year can be found in the quarterly data update report for your centre

SPAH Audit Form 2018-19

CHI Number _____

Name _____

Address _____



New Patient section



Date of diagnosis _____/_____/20____

Diagnosis Sickle Cell Disease/Thalassaemia Minor/Thalassaemia Major

Subtype _____ e.g. HBSS



New Referrals from New Born Screening only

Date family seen _____/_____/20____

NBS Lab informed

Antibiotic Prophylaxis: Prescribed Refused

Prophylaxis Date _____

Annual review section

(1st appointment after birthday)



Date seen _____/_____/20____

Diagnosis Sickle Cell Disease/Thalassaemia Minor/Thalassaemia Major

Subtype _____ e.g. HBSS

Date of next visit _____



Sickle Cell Disease

	Date	Refused
TCD date	_____	<input type="checkbox"/>

Latest Pneumovax	_____	<input type="checkbox"/>
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New treatments started

Hydroxycarbamide Date _____

_____ Date _____



Regularly Transfused Patients

Transfusion: Date _____
 Regular transfusion programme

Chelator Therapy: Date _____

Deferasirox

Desferrioxamine

Deferiprone

Ferriscan/MRI Date _____