SPAH
Scottish Paediatric and Adult Haemoglobinopathies Network

Annual Report
2022/23

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Introduction

Background

The term ‘haemoglobinopathy’ covers a range of inherited blood conditions in which haemoglobin (the oxygen carrying protein in red blood cells) is either qualitatively or quantitatively abnormal. The two main disease groups are Sickle Cell Disease (SCD) and Thalassaemia. These are lifelong genetic disorders that often result in complex medical problems.

The Scottish Paediatric and Adult Haemoglobinopathies Network (SPAH) has a remit to ensure that equitable, high quality care is delivered promptly to patients with haemoglobinopathies at all points in their journey, by a multidisciplinary health care team with knowledge of the condition. This includes minimising the risk of infections by immunisation and prophylaxis, management of drug therapies, transfusion needs and consequent iron overload to improve long-term health. Patient and parent education is also important to minimise the occurrence of sickle cell acute complications and managing these at home, where possible, thus reducing disruption to education and employment.

Due to the complex nature of Sickle Cell Disease and Thalassaemia early involvement of the specialist Haematology team is crucial to ensuring good patient outcomes. The network connects the various points of service delivery in the patient pathway and supports clinicians to work together effectively. Equity of care is supported through the use of standard guidelines and networking amongst the clinicians to share best practice.

Lead Clinician reflections

As the NHS emerged from the covid crisis, there have been inevitable clinician capacity issues which have impacted network progress is some areas. Despite these, I think the Network team and members should be congratulated in what has been achieved this year. We have continued with development of guidelines, case discussion meetings and provision of information leaflets to support patients.

Much of the new work has been aimed at laying the foundation for future projects. The mapping exercise has identified clinicians and biomedical scientists responsible in some way for the care of our patients in every Health Board in Scotland. I am sure that this piece of work will prove invaluable in years to come, serving as a route for sharing information, and supporting healthcare teams in a multitude of ways.

Part 1 of the Transition project, looking at the process as patients move from paediatric care to adult care has identified a need to support and share good practice, as well as improve some areas. This will be taken forward next year.

A huge body of work has been done on the IT side of things to ensure that our data is robust and some changes have been made with detail within the body of report. Many thanks to Hugh O'Pray and the clinicians for their support with this – it is clear to me that accurate data is the foundation to both service development and auditing of performance.

I look forward to meeting many of our patients and families at our patient event to be held later in the year. This will be an opportunity for engagement and education, and also to identify patient needs, which will help develop our future work plan.

Finally, I think it is important to end on an optimistic note and to mention the emerging therapies for patients with haemoglobinopathies. Some have not provided the clinical breakthrough that was initially hoped, but others such as gene therapy are potentially on the horizon, and we will work as a network to ensure that if and when they become available, our patients have appropriate access to them.

Current position

The majority of targets within the workplan have been delivered, albeit in some cases with extended timescales due to staff capacity, and this report gives an overview of progress.

95% of Business as Usual objectives were achieved
The patient and families event was postponed until September 2023 partly due to covid concerns regarding this patient group as well as to allow feedback to be obtained from stakeholders around the planning for the event.

89% of Service Development Plan objectives were achieved.
The objective to explore benchmarking opportunities with the rest of the UK and develop a benchmarking plan was not progressed due to incomplete SPAH KPI data collection.

Highlights

Scottish Ambulance Service & Improvements to Pre-Hospital Care

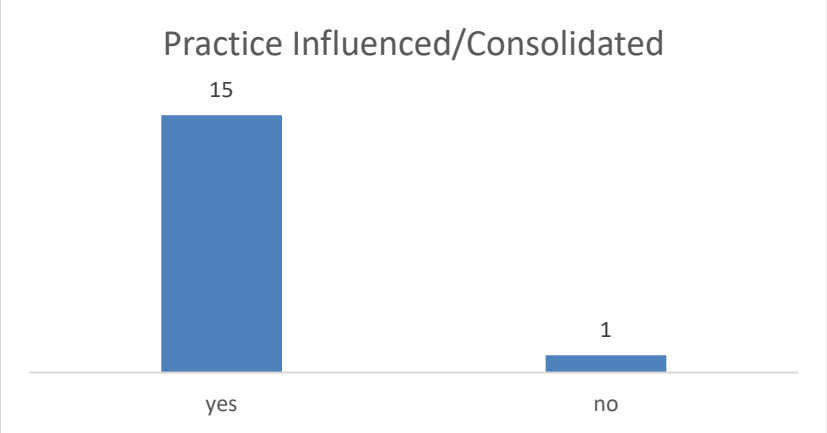
The network has completed its collaborative programme of work with the Scottish Ambulance Service with the development of a handheld patient alert card containing key information about their condition and how it should be managed during an acute crisis. A patient information leaflet on how to call an ambulance to highlight the needs of a patient experiencing a sickle cell crisis has also been produced in association with the Scottish Ambulance Service and is available on the SPAH Website. The network plans to re-audit patients' experience of using the Scottish Ambulance Service during a sickle cell crisis to identify if there has been improvement in this area (2024/25 workplan).

Climate Sustainability

The network continued to make use of technology and remote communications to progress work this year. This has continued to be effective, saving time on travel and promoting economic and climate friendly practices.

Education

The network hosted a successful virtual education event for Haematology Registrars and trainees in September 2022. Around 30 people attended each of the sessions held on 14th and 21st September and evaluations were received from 15 participants. 94% of respondents indicated that their practice was influenced or consolidated following the events. The attendees are Scotland's consultants of the future, who are likely to end up working in all areas of Scotland (and elsewhere). It is therefore anticipated that this event has widened knowledge of the care of patients with haemoglobinopathies across Scotland.



An education channel has been created using MS Teams hosting recordings from the event to allow attendees to revisit learning at their own convenience.

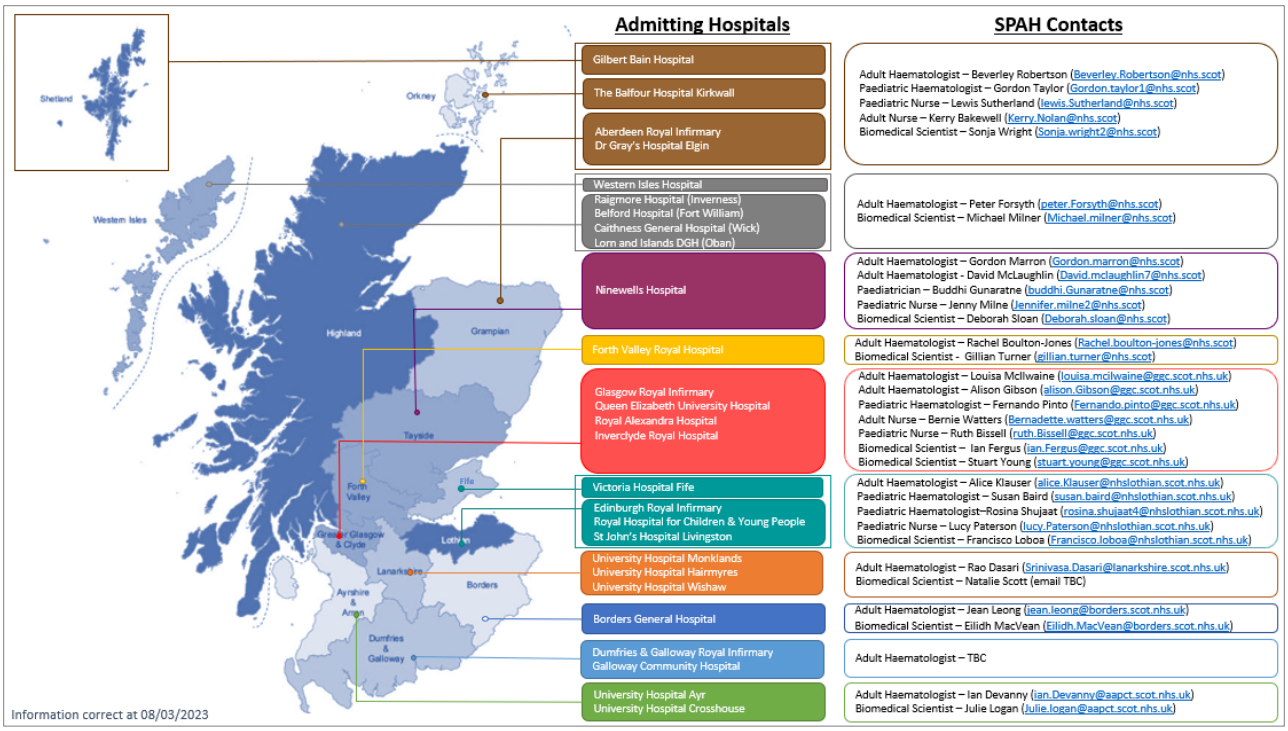
Case Discussion Meetings
Four meetings took place during the year and 22 cases were presented. In addition to case presentations with learning points this meeting allows specialist peer support. These meetings will be evaluated during the course of 2023/24.

Education Strategy
To inform the SPAH Education Strategy, a SPAH Education Working Group will be set up to review the education, training and leadership, and practice development requirements of its stakeholders. This group will also lead on the planning for a patient and family's event that is scheduled for Autumn 2023.

Mapping Exercise
A survey was shared with Scottish Haematologists through the Scottish Haematology Society in March 2022. The purpose of the questionnaire was to collect information on haematology services across Scotland to develop a dataset of professionals caring for patients with haemoglobinopathies, linked hospitals and/or treatment centres and how those centres currently access SPAH information and guidance.

The questionnaire was shared with approximately 360 professionals through the Scottish Haematology Society, 25 responses were received each providing either individual clinician, whole department, or health board information.

Feedback was used to create a SPAH service map highlighting each health board, linked hospitals and the associated haematology clinicians.



The network took the opportunity during the mapping exercise to ask clinicians if they used the guidelines available on the SPAH website and 69% of those who responded confirmed that they did. All the clinicians currently using guidelines identified that they found the guidelines helpful or somewhat helpful.

The deliverables of the mapping project will provide benefits to patients, services and the SPAH network including:

- Improved access to education including Scottish case discussion meetings and West London HCC MDTs
- SPAH guidelines and updates will be shared widely across NHS Scotland through accurate clinician lists and additional distribution routes
- Accurate data and audit through additional boards reporting against KPIs
- Accuracy of patients with haemoglobinopathies registered on CAS
- Identification of key personnel in every Health Board area who may be able to support future projects.

Review of Transition Project – Phase 1

The SPAH Steering Group identified patients moving from paediatric to adult services as an area for review. A questionnaire was circulated in 2018 and a small amount of feedback was received. Due to the very small number of patients transferring between services the project lost momentum. It was agreed to refresh the project using project management and improvement methodology.

To garner as much information as possible for a comprehensive understanding of current transition practices in haemoglobinopathy services across NHS Scotland, the SPAH Programme Manager and NSS Assistant Programme Manager met (via MS Teams) with healthcare professionals from NHS Boards looking after patients with haemoglobinopathies.

Common and key responses were noted throughout the information gathering which are summarised below.

Current practice:

- processes are in place in all areas for transition (preparing for transfer) and transfer (handover from paediatric to adult services
- the process is mostly informal with services being aware that the Ready Steady Go Hello programme is available to access to support the process
- the age at which the transition process starts ranges from 11-15 years, and is decided by the local service

Challenges:

- there are difficulties around children over the age of 16, who have not transferred to adult services, being admitted to A&E or via acute medicine as they are expected to be managed in the adult sector
- there is capacity and resource issues across paediatric and adult services
- there is lack of familiarity of managing haemoglobinopathy patients in some adult hospitals

Key to understanding the effectiveness of current transition practices are the views and experiences of patients. A questionnaire was developed and circulated to adult haematology services and via The Hope Project Scotland to share with patients who had transferred from paediatric to adult services within the last five years, however only 2 responses were received. This was identified as a risk at the beginning of the project.

The SPAH Steering Group considered the data collected during Phase 1 of the project and agreed to set up a Short Life Working Group to progress the development of good practice guidelines.

The information gathered by SPAH will feed into a separate National Network Management Service project which is scoping transition across all paediatric networks in Scotland.

Patients with haemoglobinopathies registered on the National Clinical Audit System

Patient demographics

The Clinical Audit System (CAS) continues to be the national register for SPAH. There are a total of 406 active follow up patients currently registered on CAS, an increase of 65 patients from 2021/22. In almost all Health Boards the patient numbers are rising, and within the last 5 years, the number of patients in Scotland with a haemoglobinopathy diagnosis has increased by 87.96%. The network will review the impact these increases in patient numbers are having on local services to understand what SPAH can do to support this.

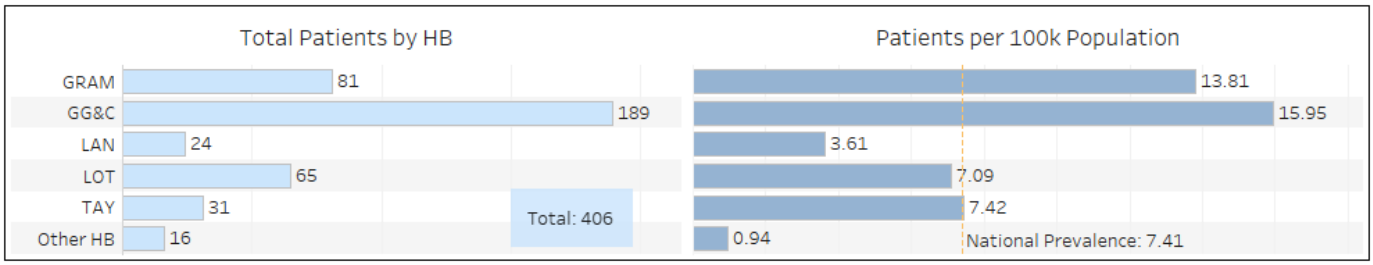
a) Total number of patients by Health Board

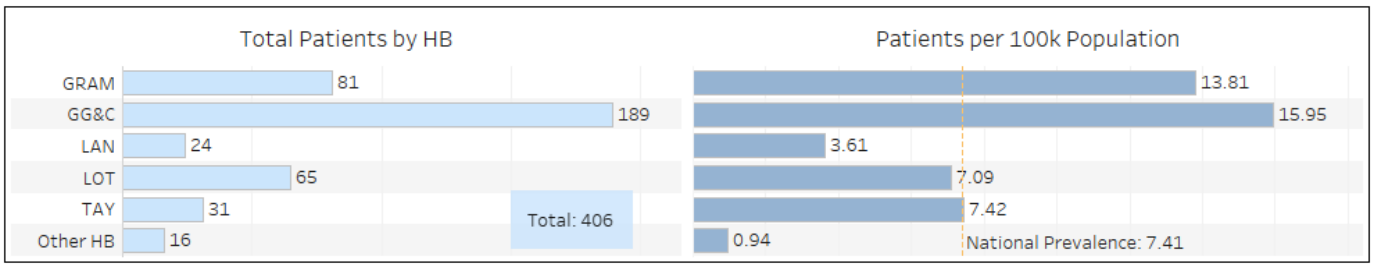
Total Patients by Health Board

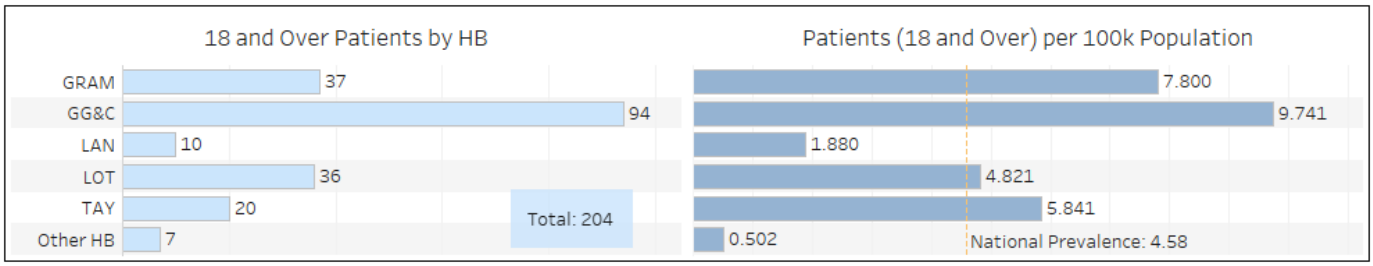
	17/18	18/19	19/20	20/21	21/22	22/23	% Change Since 17/18
Scotland	216	231	290	313	341	406	87.96%
GRAM	39	37	47	55	56	81	107.69%
GG&C	98	112	148	157	177	189	92.86%
LAN	10	10	15	19	20	24	140.00%
LOT	41	41	49	51	55	65	58.54%
TAY	17	20	23	24	23	31	82.35%
Other HB	11	11	8	7	10	16	45.45%

b) Adult & paediatric population per 100K population

The paediatric and adult patients registered on CAS by Health Board of Residence and by 100k of Head of Population is shown in the next three slides. This data is likely to have many uses, but in particular highlights the number of patients cared for by the tertiary paediatric centres who will be returning to the local health boards when they reach adulthood. (Paediatric patients are cared for in one of 4 tertiary paediatric centres, but adult care is provided in all healthboards of residence).







c) Specific disease data by treatment centre

The largest group is patients with sickle cell disease, with the distribution across Scotland shown in the tables below:

Patients and Conditions by Treatment Centre

	Sickle Cell Disease	Thalassaemia Intermedia	Thalassaemia Major
A&A - Adults	<5		
Aberdeen - Adults	32	<5	<5
Aberdeen - Children	38	<5	
Dundee - Adults	16	<5	<5
Dundee - Children	<5	<5	<5
Edinburgh - Adults	35	<5	<5
Edinburgh - Children	19	<5	6
Forth Valley	<5		
Glasgow - Adults	85	8	<5
Glasgow - Children	90	15	12
Lanarkshire - Adults	6		

Figures in these graphics for previous years may not match those published in previous annual reports. The figures for previous years have been refreshed using the list of active patients for each year combined with the current clinical data and the current method of calculating the KPI's.

The figures have been refreshed for several reasons, including:

1. Data collection is an ongoing process and a review of CAS data has found that improvements in data recording since original publication has led to significant changes in KPI achievement.
2. The calculation method for some KPI's has been changed this year (KPI 4.2 now includes patients aged 2-7 and KPI 7 now accepts reviews in the last 15 months). Refreshing the figures for previous years using the new calculation method allows a like for like comparison of achievement.
3. It was agreed to change the reporting period for KPI's 1 and 2 from financial year to calendar year to bring them into line with KPI's 3 and 4. Refreshing the figures for previous years allows us to present the data without gaps or overlapping reporting periods.

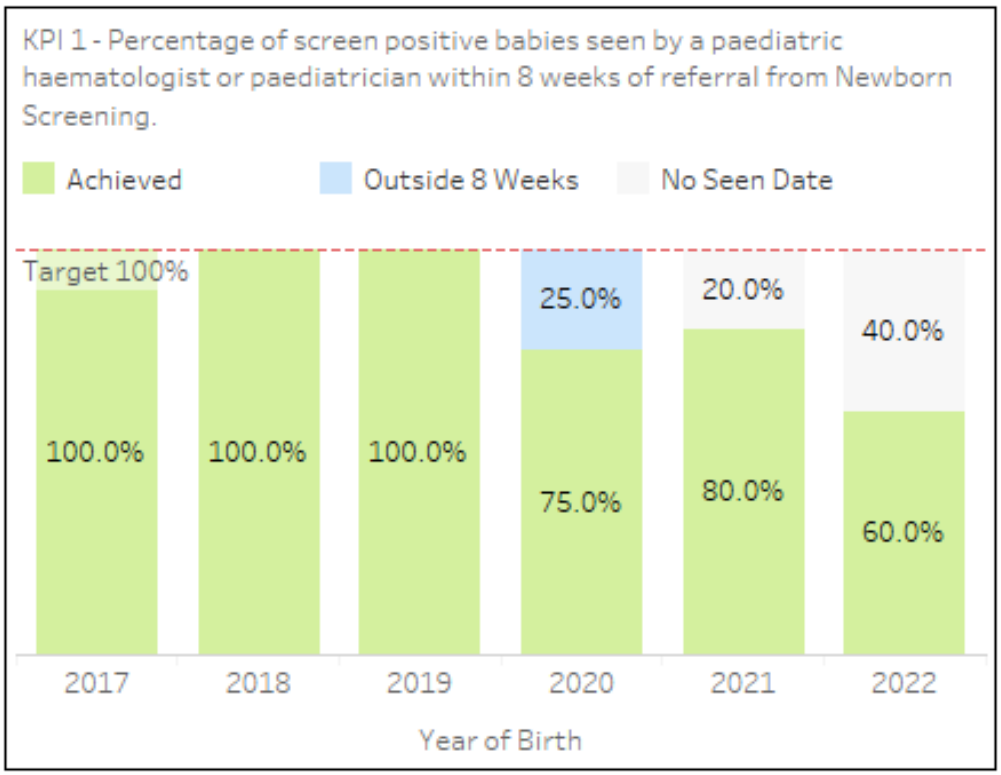
The refreshed achievement may go up or down. In most cases a rise can be attributed to missing data being added to the system. Indicators can fall because new patients are included in the denominator where a. the KPI has been expanded (in the case of KPI 4.2) and b. missing diagnosis data is added which in turn adds a patient to the population for a specific KPI.

Reporting Against SPAH Key Performance Indicators

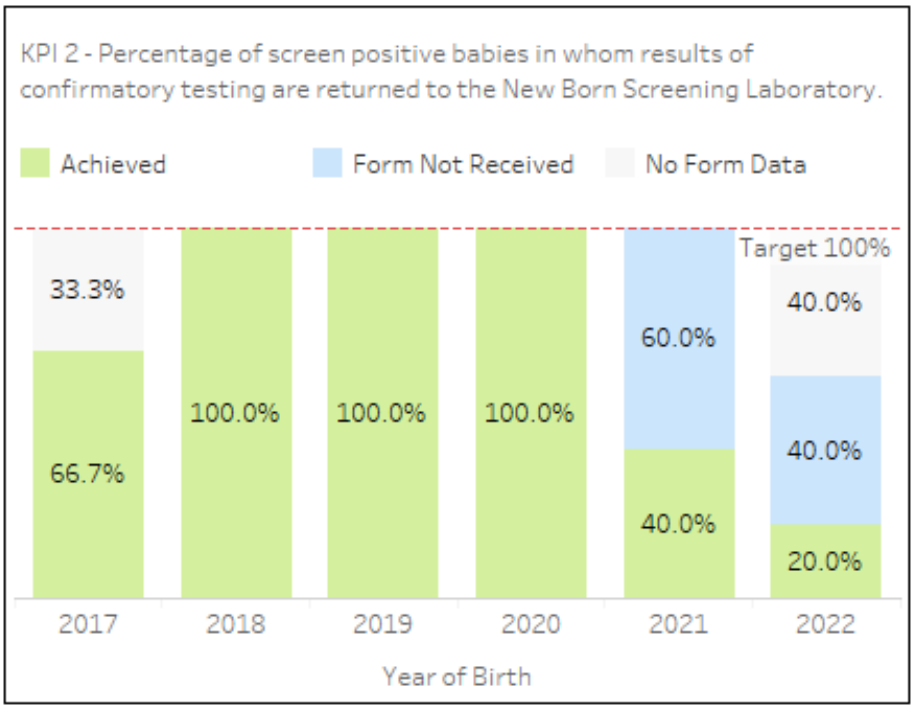
Measuring performance has once again been an objective for the network during 2022/23. Clinicians have continued to provide data to measure against 7 Key Performance Indicators (KPIs).

The data output for all the KPIs is only as good as the data put in. Clinician time and late access to the KPI reporting spreadsheetshas had a significant impact on the data input into the CAS system, beyond the basic demographics. It is recognised that this is something that needs to be addressed going forward. Clinicians have fed back their impression that most KPI activity is being undertaken however not recorded on CAS due to clinical pressures.
KPI data which is available within CAS is provided below.

KPI 1 – 100% of screen positive babies are seen by a paediatric haematologist or paediatrician within 8 weeks of referral from Newborn Screening.

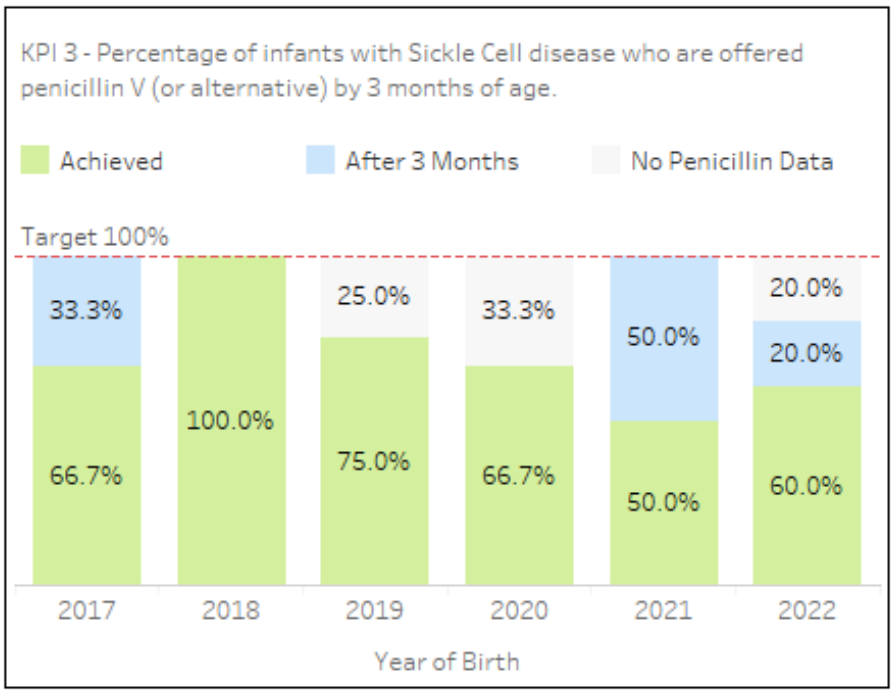


KPI 2 – 100% of screen positive babies in whom results of confirmatory testing are returned to the New Born Screening Laboratory.

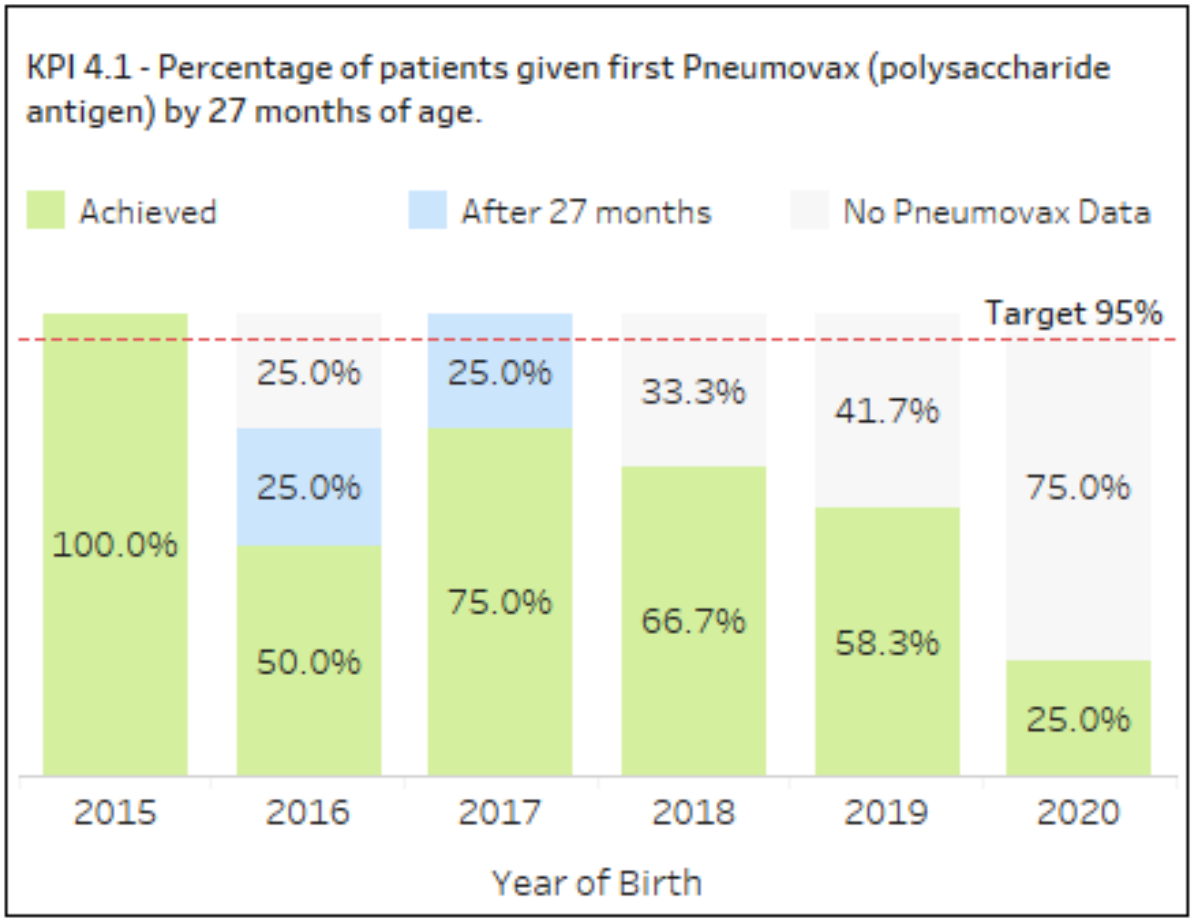


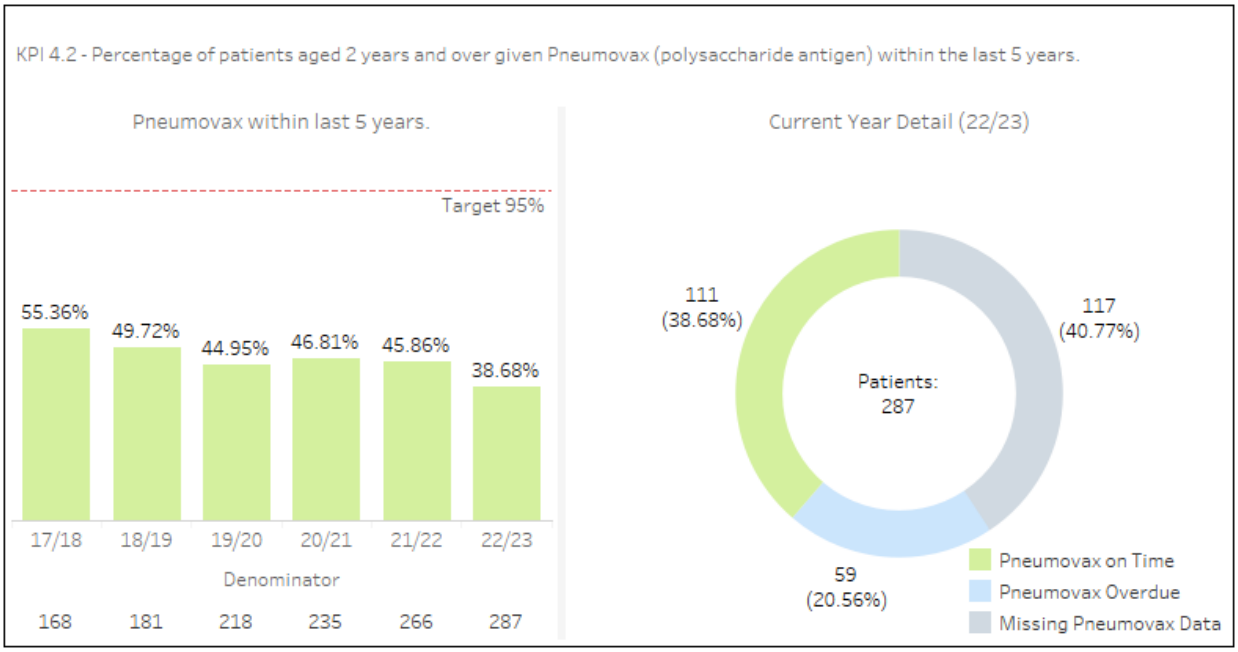
NB "Form not received" relates to the Newborn Screening Service not receiving paperwork from clinicians highlighting confirmatory testing results.

KPI 3 - 100% of patients with sickle cell disease are offered penicillin V (or alternative) by 3 months of age.



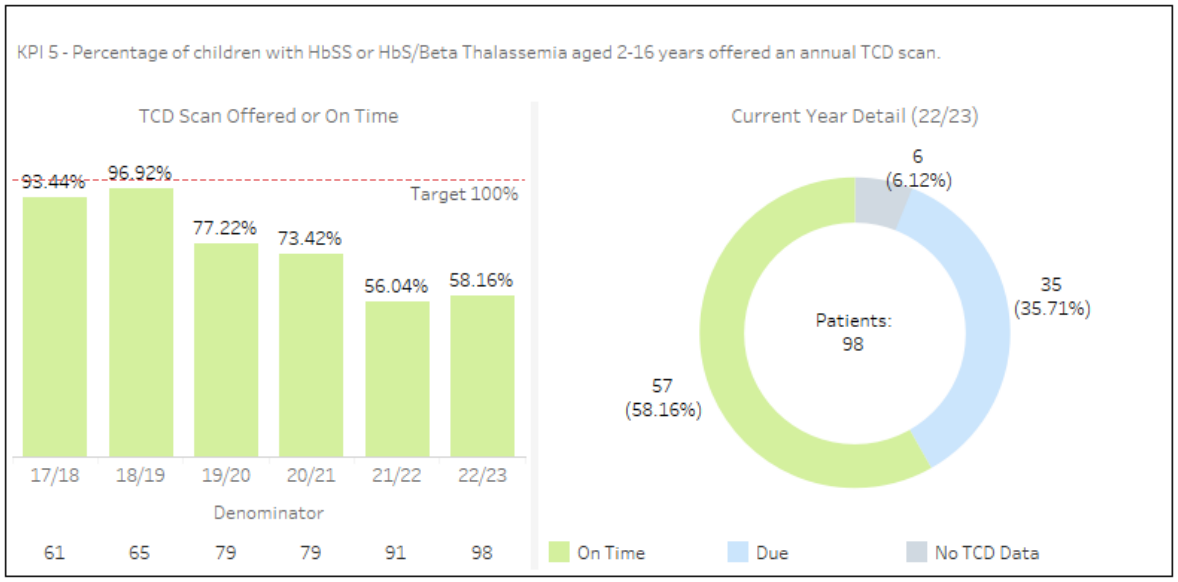
KPI 4 – 95% of patients should be given first Pneumovax (polycaccharide antigen) by 27 months and 5 yearly thereafter.



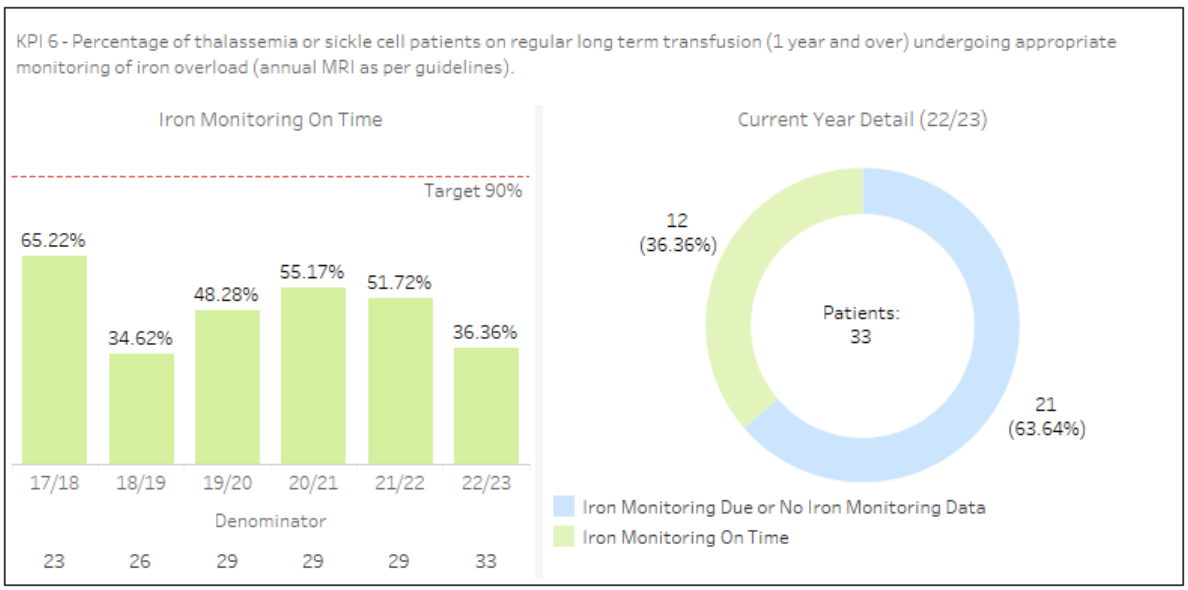


KPI 5 – 100% of children with HbSS or HbS/Beta thalassemia aged 2-16 years <u>offered</u> an annual TCD scan.

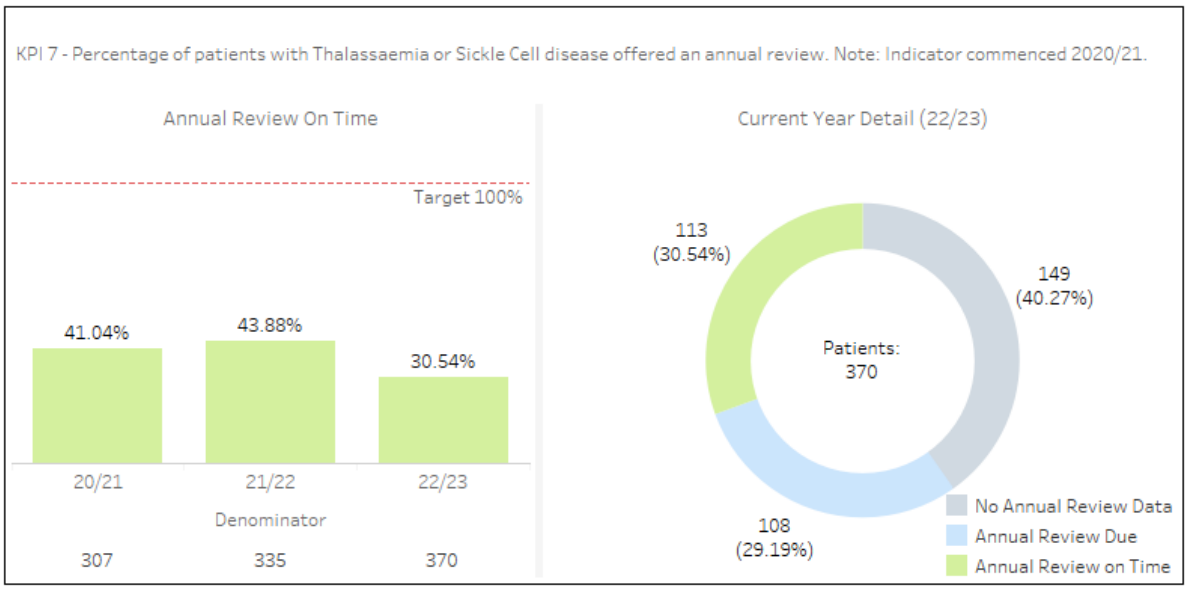
Transcranial Doppler scanning was limited during the pandemic and the service has been struggling to catch up since. Patients at highest risk of abnormal scans requiring intervention have been prioritised, although it is recognised that there is a way to go to catch up to pre pandemic screening levels.



KPI 6 – 90% of thalassemia patients on regular transfusion undergoing appropriate monitoring of iron overload (annual MRI) as per guidelines. MRI scan within the last 12 months.



KPI 7 – 100% of patients with Thalassaemia or Sickle Cell disease should be offered an annual review.



Note: the denominator this year is 370 (rather than the 373 in the first set of graphics) because 3 patients are under 1 years old and so are not included.

As commented above, the data coming out is only as good as that going in. This year we have prioritised capturing accurate demographic data. Improving data collection needs to be addressed going forward, and there is a plan to task the Audit & Data Working Group with doing so. In the absence of data managers for each Health Board, what is likely needed is identification of a limited number of KPIs to focus on, which can be added to over time, as the current data capture is overwhelming. There is also the opportunity to review whether we need to change any of the KPIs/add different ones.

Looking forward – 2023/24

Over the course of 2023/24 the programme of work will include:

- Phase 2 – Transition Project
- Parent/Family Event
- Future provision of Adult Ferriscan
- SPAH response to the No One's Listening Report
- Development of 3-5 year Strategic Plan
- Comprehensive review of website content
- SPAH Education Working Group to be set up to review the education, training and leadership, and practice development requirements of its stakeholders.
- Audit linkage between antenatal screening, newborn screening and paediatrics
- Explore benchmarking opportunities with another UK treatment centre.
- Review of guidelines/protocols & pathways
- Development of Patient Forum
- Case discussion meeting management
- Review current audit and data timetable

Risks and issues

Adult Ferriscan Service

The Ferriscan service was previously provided to all adult patients at risk of transfusion related iron overload in Scotland at two locations (Grampian and Forth Valley). When these services ceased, to mitigate the impact of Ferriscan being unavailable a short-term pathway was developed supporting referral of adult patients for scanning at the Queen Elizabeth University Hospital in Glasgow which commenced in May 2021.

NHS Lothian Radiology Department commenced the provision of FerriScan for all the iron assessment MRIs for GI/liver and haematology patients in January 2023.

Funding is in place for all Scottish patients for the next year for 2023/24, however with the Lothian service now undertaking their own scans the current model no longer provides a national service.

The network will explore the feasibility of scans being provided more locally in NHS Boards although recognises that if this is not possible then a solution will be required to ensure equity of access. Funding is a separate but related issue and will also need explored.

Guideline Review

The network has a timetable for the review and development of guidelines. The following documents (16 guidelines, 2 pathways and 4 leaflets) have been reviewed during 2022/23.

- Paediatric & Adult Guideline - Physiotherapy - Acute Management of Sickle Cell Disease
- Paediatric & Adult Guideline - Introduction to Sickle Cell Disease for Physiotherapists
- Paediatric & Adult Guideline - Hyperhaemolysis
- Paediatric Guideline - Iron overload and chelation therapy
- Paediatric Guideline - Acute Chest Syndrome
- Paediatric Guideline - Diagnosis and Management of Non-Transfusion-Dependent
- Thalassaemia phenotypes
- Paediatric Guideline - Use of Hydroxycarbamide in Sickle Cell Disease
- Paediatric Guideline – The Management of Acute Abdominal Pain in Children with Sickle Cell Disease
- Paediatric Guideline – Inpatient Specialist management of acutely unwell children with haemoglobinopathies
- Paediatric Guideline – Painful Sickle Cell Crisis (assessment and management)
- Paediatric Guideline – Stroke and other CNS Manifestations in Sickle Cell Disease

- Adult Guideline - Use of Iron Chelation in Sickle Cell Disease & Thalassaemia
- Adult Guideline - Acute Chest Syndrome in Sickle Cell Disease
- Adult Guideline - Perioperative management of adult patients with Sickle Cell Disease
- Adult Guideline – Stroke and other CNS complications in sickling disorders
- Adult Guideline - Thalassaemia Outpatient Management

- Managing Iron Overload Information Leaflet
- Managing Acute Chest Syndrome Information Leaflet Iron Overload Information Leaflet
- Calling an Ambulance Information Leaflet
- Caring for Unwell Child at Home Information Leaflet

- Patient Pathway - Sickle Cell Disease or Thalassaemia diagnosed outwith Newborn Screening
- Patient Pathway - New case identified by Newborn Screening

There has been a delay in the review of 6 network guidelines and 10 patient leaflets due to the impact of the covid pandemic compounded by staff capacity. Overdue guidelines have been risk assessed by sub-group chairs to confirm that they can remain on the SPAH website.

The Steering Group have discussed possible solutions to manage such a large number of documents including:

- Document management update reports at Steering Group Meetings (already implemented)
- Combining guidelines to have two overarching documents (one for all Paediatrics and one for all Adult guidelines)
- One year extension for guidelines that do not require any changes (agreed by Chair of Sub-Group)
- Reviewing some guidelines at steering group meetings.

These solutions will be explored further at the Paediatric and Adult Guidelines Sub-Group meetings. Guidelines are available from the SPAH website - http://www.spah.scot.nhs.uk/professionals/

Finance

The network has spent £1,133 of its £5,000 budget during 2022/23.

This included printing costs for the alert cards and school's booklet as well as costs for Plain English review of network leaflets.