SPAH
Scottish Paediatric and Adult Haemoglobinopathies Network

Annual Report
2023/24

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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.

Current position

The majority of targets within the workplan have been delivered and this report gives an overview of progress.

24/26 (92%) of Business as Usual objectives were achieved
Two subgroups had one meeting less than planned in 2023/24 due to capacity.
It was agreed that the objective to engage with patients and families through delivery of a patient event could be delayed due to financial pressures. It is hoped this will be able to progress in 2024/25.

8/10 (80%) of Service Development Plan objectives were achieved.
Progressing the development of the transition and transfer guidance was delayed due to nursing capacity and will be completed in 2024/25.
An education group has not yet been established though education was considered as part of the strategic planning event in January 2024.

Highlights

Strategy session

The network held a strategy session in Perth in January 2024 to develop a long term vision and a strategic plan setting out how the network will work towards that vision over the next few years. To inform this work, a survey was undertaken to identify what mattered to patients.

Patients told us:

- The care, availability, knowledge and relationships with the multidisciplinary specialist clinicians were appreciated by them.
- They valued being seen regularly with no waiting list.
- There were challenges with knowledge and awareness in the wider clinical workforce.
- They felt that access to services and the experience of emergency care could be improved.

The detailed information from patients informed the session along with the findings and recommendations from the “No-one’s Listening” report. The session focused on three key areas:

- Paediatrics
- Adult outpatient management
- Adult acute care

The session generated a number of improvement ideas which will be developed into a strategic plan. The priority areas form part of the 2024/25 workplan.

Antenatal Screening, Newborn Screening and Paediatrics Audit

The purpose of this audit was to identify if sickle screen positive babies in Scotland have been
correctly
identified as

high-risk pregnancy by the antenatal screening programme and that this information was then correctly communicated to the Newborn Screening (NBS) laboratory followed by timely referral to the appropriate paediatric sickle centre. Review of this process is to identify if key performance indicators are being met and if not, how the process can be improved.

Although the data was incomplete, a number of improvement areas were identified and discussed. The paper was shared with the Senior Programme Manager for screening in NSD and it was agreed that as an initial next step local teams should look at data for their area in more detail.

Case Discussion Meetings

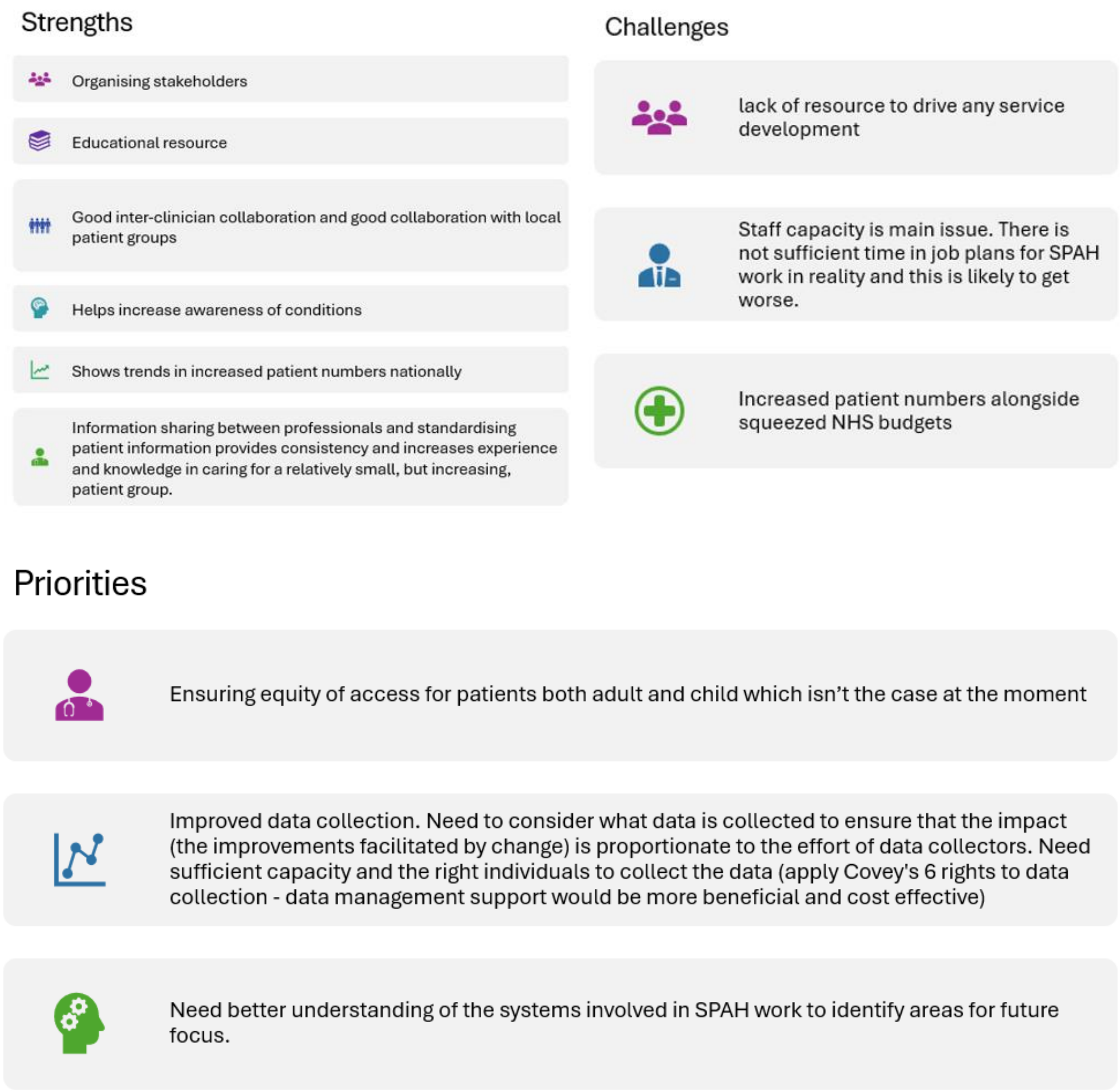
SPAH continued to facilitate peer support for specialists through delivery of 6 case discussion meetings using Teams. There were 23 cases and 3 adverse events discussed across the year with an average attendance at events of 10 people. This format supports patient care and allows sharing of clinical knowledge.

Stakeholder Survey

A stakeholder survey was shared in March 2024 to gather feedback from people who currently work within, are involved in, or are impacted by the network. There were 8 responses and a lot of positive feedback was received and highlights the value added by the network:

- 100% responses ‘strongly agreed’ or ‘agreed’ that the network makes a positive difference to patients.
- 100% responses ‘strongly agreed’ or ‘agreed’ that the network makes a positive difference to staff.
- 87% responses ‘strongly agreed’ or ‘agreed’ that the network provided a structure to make service improvements in haemoglobinopathies care.
- 87% responses ‘strongly agreed’ or ‘agreed’ that the networks service development activity adds value to haemoglobinopathies care.
- 100% of applicable responses ‘strongly agreed’ or ‘agreed’ that the networks education offering adds value to haemoglobinopathies care.
- 63% responses ‘strongly agreed’ or ‘agreed’ that the networks audit and continuous quality improvement activity adds value to haemoglobinopathies care.

The survey was also an opportunity for stakeholders to share feedback on network strengths, network challenges, areas for improvement and the priorities for the next few years. The responses are summarised below:



Strengths

- Organising stakeholders
- Educational resource
- Good inter-clinician collaboration and good collaboration with local patient groups
- Helps increase awareness of conditions
- Shows trends in increased patient numbers nationally
- Information sharing between professionals and standardising patient information provides consistency and increases experience and knowledge in caring for a relatively small, but increasing, patient group.

Challenges

- lack of resource to drive any service development
- Staff capacity is main issue. There is not sufficient time in job plans for SPAH work in reality and this is likely to get worse.
- Increased patient numbers alongside squeezed NHS budgets

Priorities

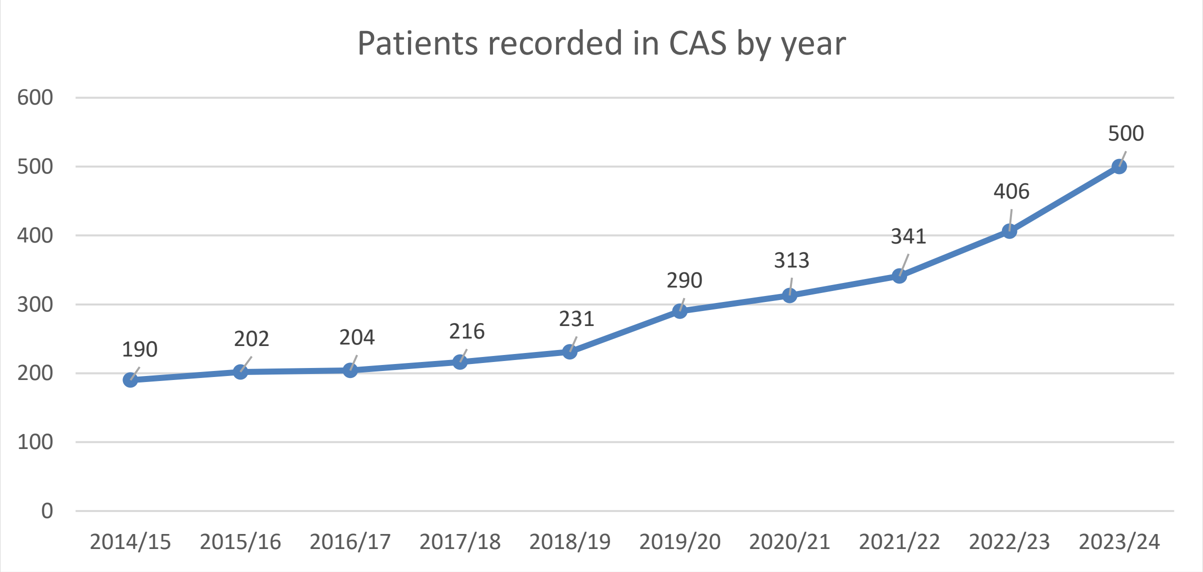
- Ensuring equity of access for patients both adult and child which isn't the case at the moment
- Improved data collection. Need to consider what data is collected to ensure that the impact (the improvements facilitated by change) is proportionate to the effort of data collectors. Need sufficient capacity and the right individuals to collect the data (apply Covey's 6 rights to data collection - data management support would be more beneficial and cost effective)
- Need better understanding of the systems involved in SPAH work to identify areas for future focus.

The network will consider the full results of the survey and identify any actions.

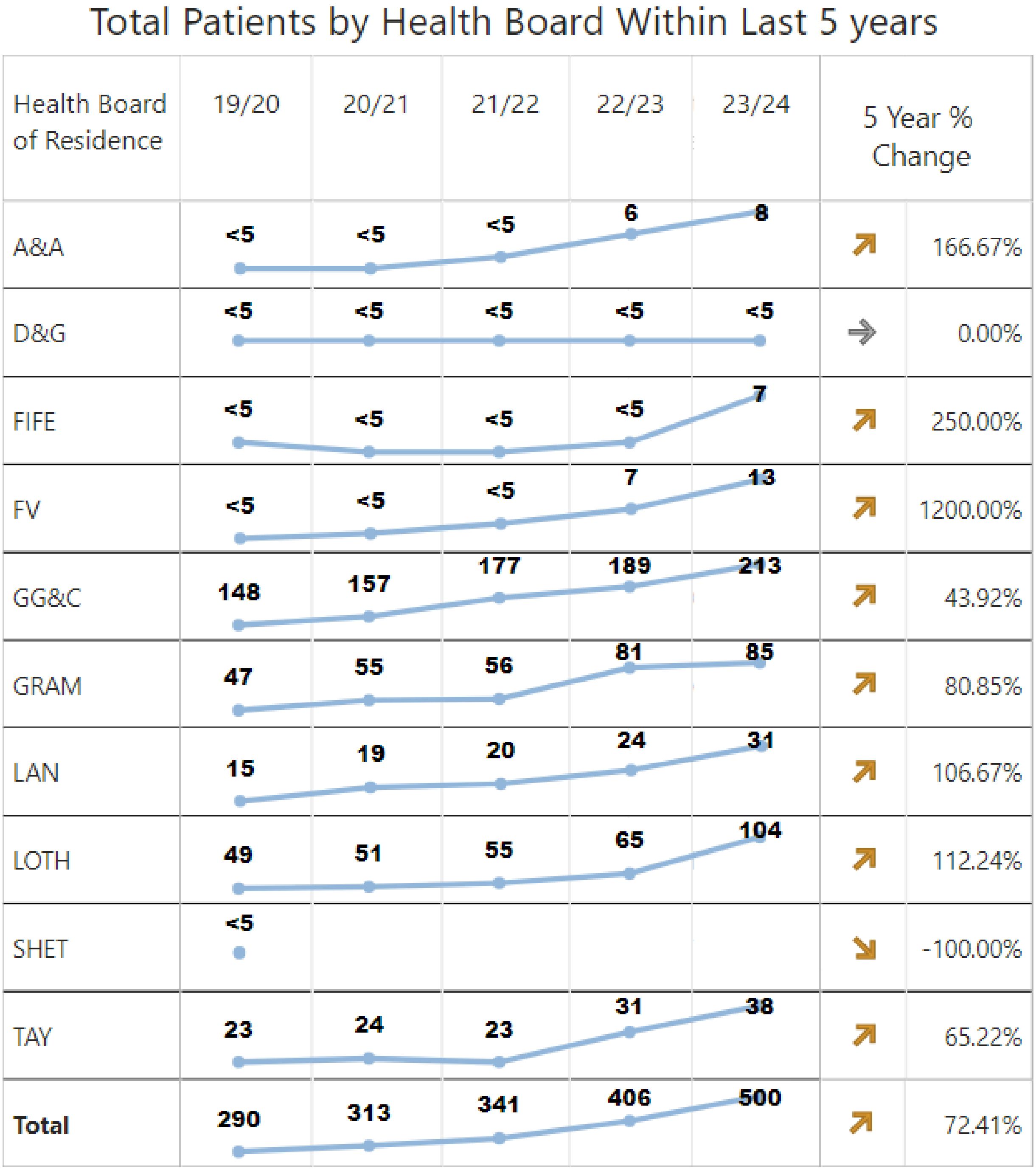
Patients with haemoglobinopathies registered on the National Clinical Audit System

Patient demographics

The number of patients with haemoglobinopathies in Scotland has risen significantly in the last 10 years, creating considerable pressure on services. Since 2014 patient numbers have increased by over 150%, more than doubling in the last 5 years alone, and continue to rise. It is clear that this is predominantly a genuine rise in numbers rather than more robust data collection. There are also now a number of health boards outside of the main centres that have small but rising numbers of patients. In recognition of these trends, SPAH have identified the need to review the model of care as part of the strategic work plan. This is scheduled to begin in 2025/26.



a) Total number of patients by Health Board

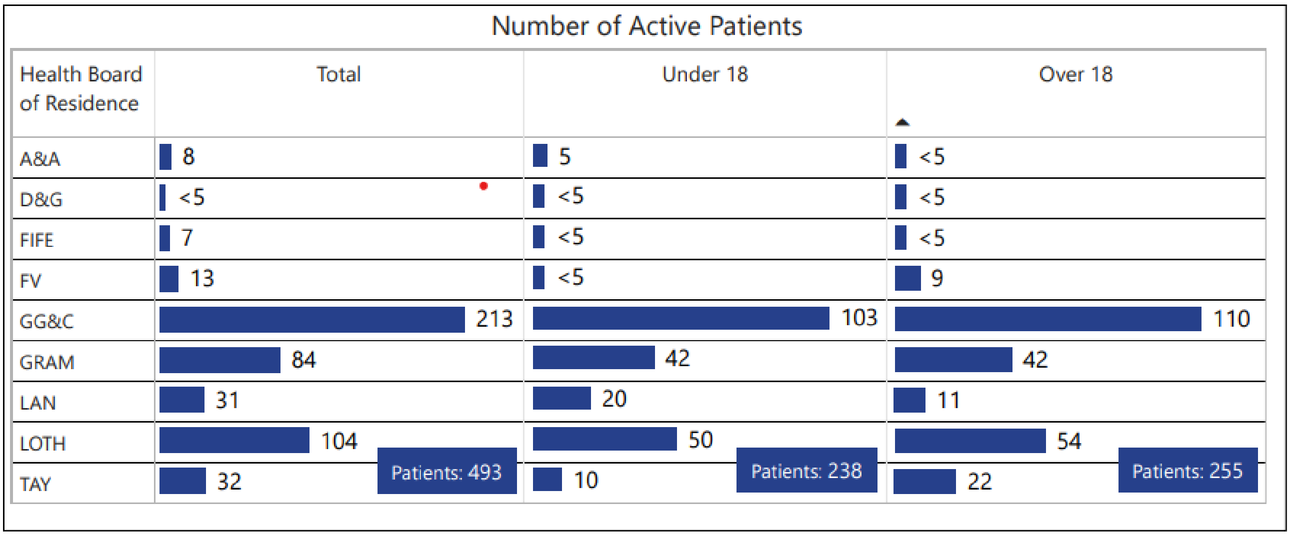


Total Patients by Health Board Within Last 5 years

Health Board of Residence	19/20	20/21	21/22	22/23	23/24	5 Year % Change	
A&A	<5	<5	<5	6	8	↗	166.67%
D&G	<5	<5	<5	<5	<5	→	0.00%
FIFE	<5	<5	<5	<5	7	↗	250.00%
FV	<5	<5	<5	7	13	↗	1200.00%
GG&C	148	157	177	189	213	↗	43.92%
GRAM	47	55	56	81	85	↗	80.85%
LAN	15	19	20	24	31	↗	106.67%
LOTH	49	51	55	65	104	↗	112.24%
SHET	<5					↘	-100.00%
TAY	23	24	23	31	38	↗	65.22%
Total	290	313	341	406	500	↗	72.41%

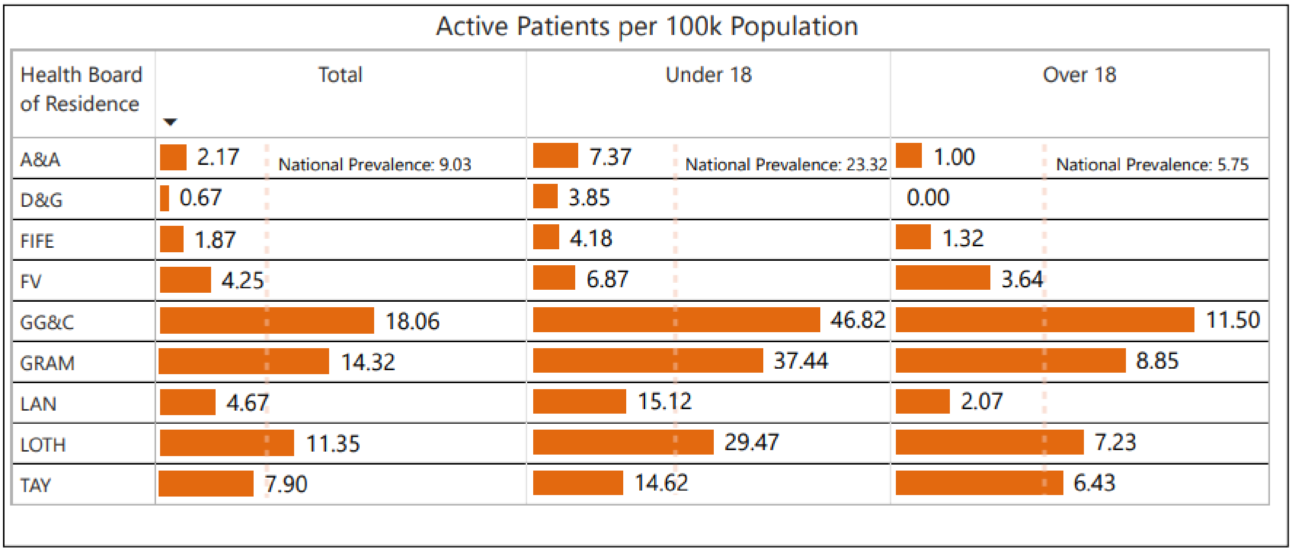
b) Adult & paediatric population per 100K population

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 two 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).



Number of Active Patients

Health Board of Residence	Total	Under 18	Over 18
A&A	8	5	<5
D&G	<5	<5	<5
FIFE	7	<5	<5
FV	13	<5	9
GG&C	213	103	110
GRAM	84	42	42
LAN	31	20	11
LOTH	104	50	54
TAY	32	10	22
	Patients: 493	Patients: 238	Patients: 255

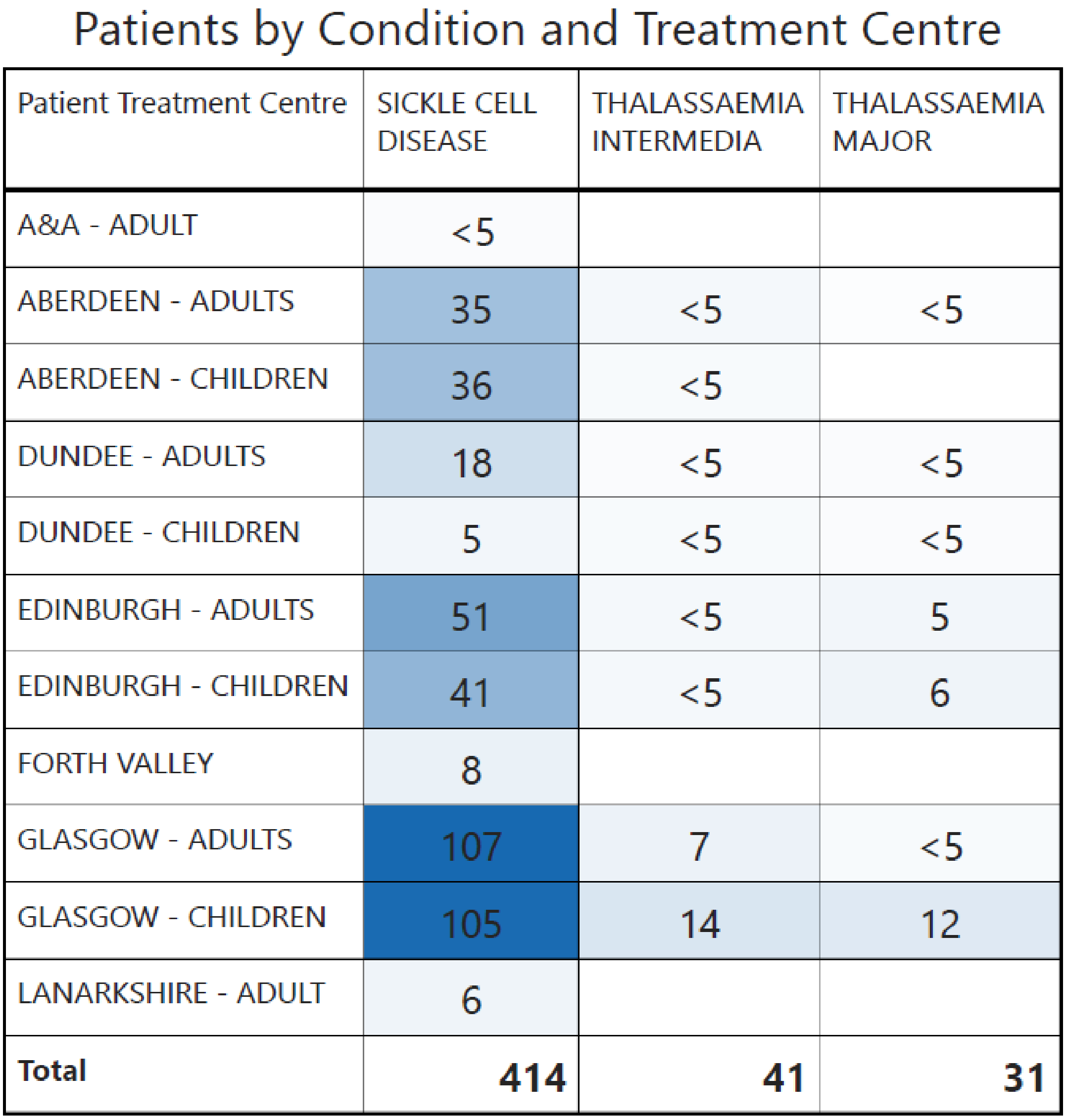


Active Patients per 100k Population

Health Board of Residence	Total	Under 18	Over 18
A&A	2.17	7.37	1.00
D&G	0.67	3.85	0.00
FIFE	1.87	4.18	1.32
FV	4.25	6.87	3.64
GG&C	18.06	46.82	11.50
GRAM	14.32	37.44	8.85
LAN	4.67	15.12	2.07
LOTH	11.35	29.47	7.23
TAY	7.90	14.62	6.43
	National Prevalence: 9.03	National Prevalence: 23.32	National Prevalence: 5.75

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 by Condition and Treatment Centre

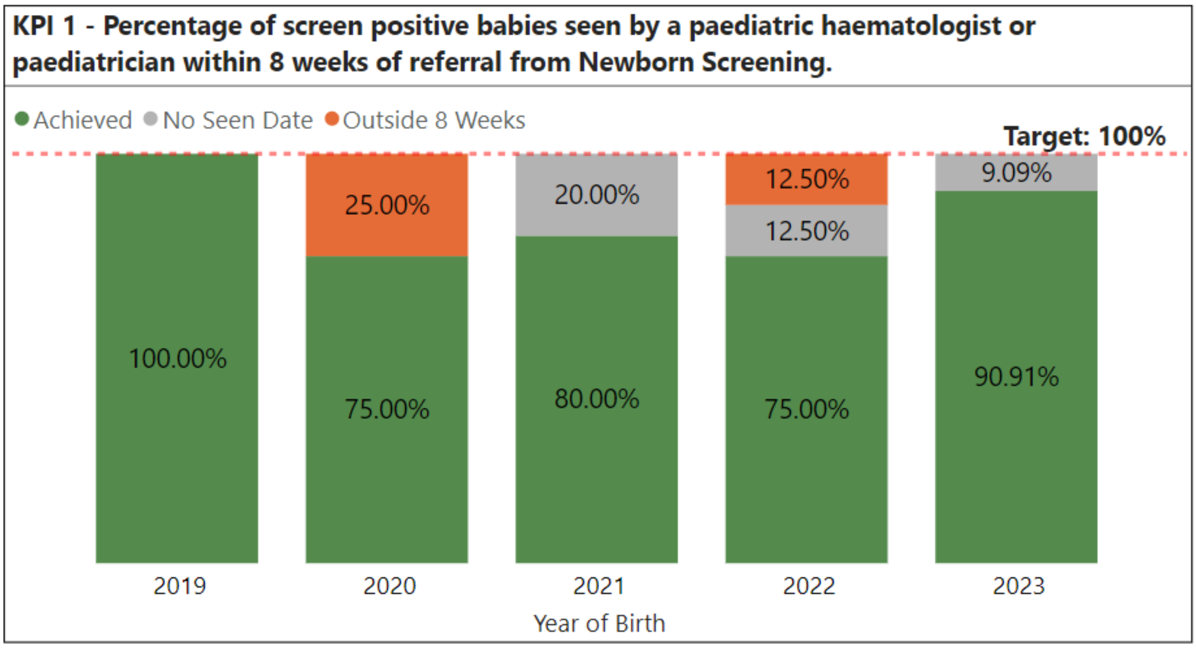
Patient Treatment Centre	SICKLE CELL DISEASE	THALASSAEMIA INTERMEDIA	THALASSAEMIA MAJOR
A&A - ADULT	<5		
ABERDEEN - ADULTS	35	<5	<5
ABERDEEN - CHILDREN	36	<5	
DUNDEE - ADULTS	18	<5	<5
DUNDEE - CHILDREN	5	<5	<5
EDINBURGH - ADULTS	51	<5	5
EDINBURGH - CHILDREN	41	<5	6
FORTH VALLEY	8		
GLASGOW - ADULTS	107	7	<5
GLASGOW - CHILDREN	105	14	12
LANARKSHIRE - ADULT	6		
Total	**414**	**41**	**31**

Reporting Against SPAH Key Performance Indicators
Measuring performance has once again been an objective for the network during 2023/24. Clinicians have continued to provide data to measure against 7 Key Performance Indicators (KPIs).

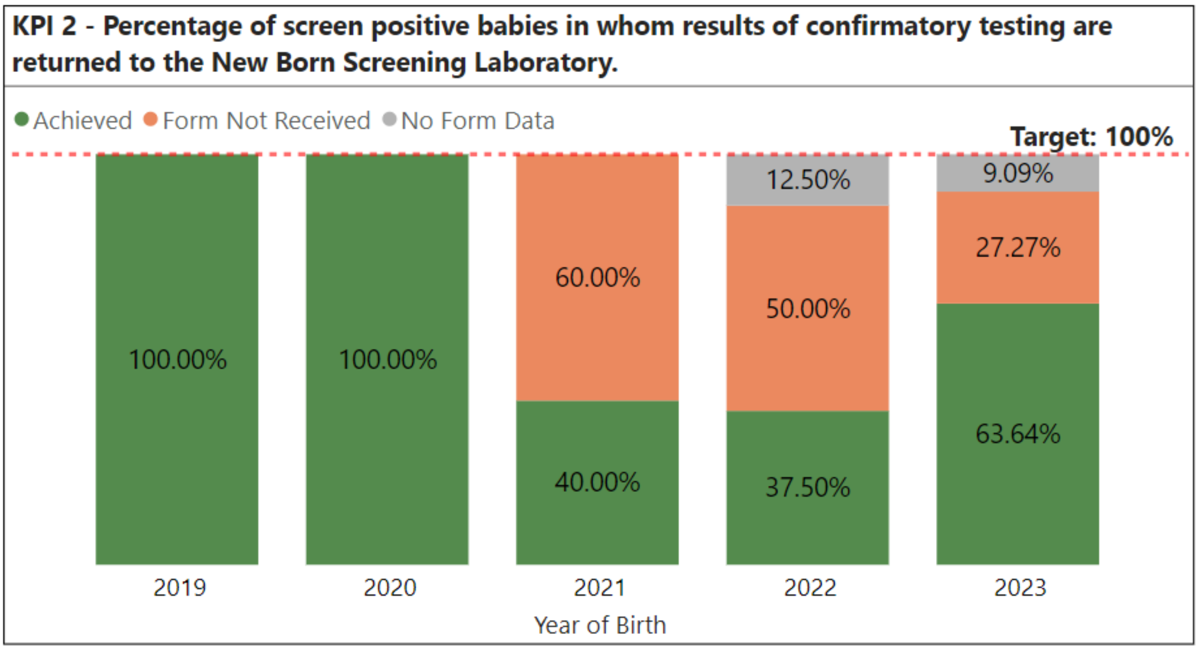
The data output for all the KPIs is only as good as the data put in. Clinician time and challenges with accessing the CAS has had a significant impact on the data input into the CAS system, beyond the basic demographics. It is recognised that this is something that needs to be addressed going forward.

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.

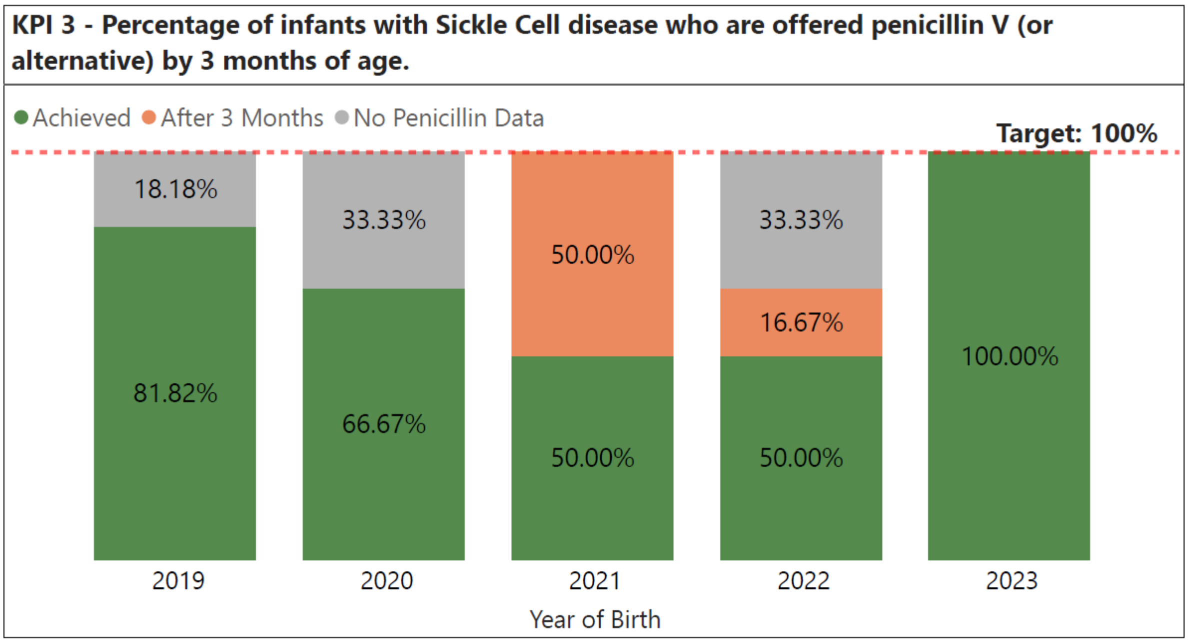


KPI 2 – 100% of screen positive babies in whom results of confirmatory testing are returned to the New Born Screening Laboratory.



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

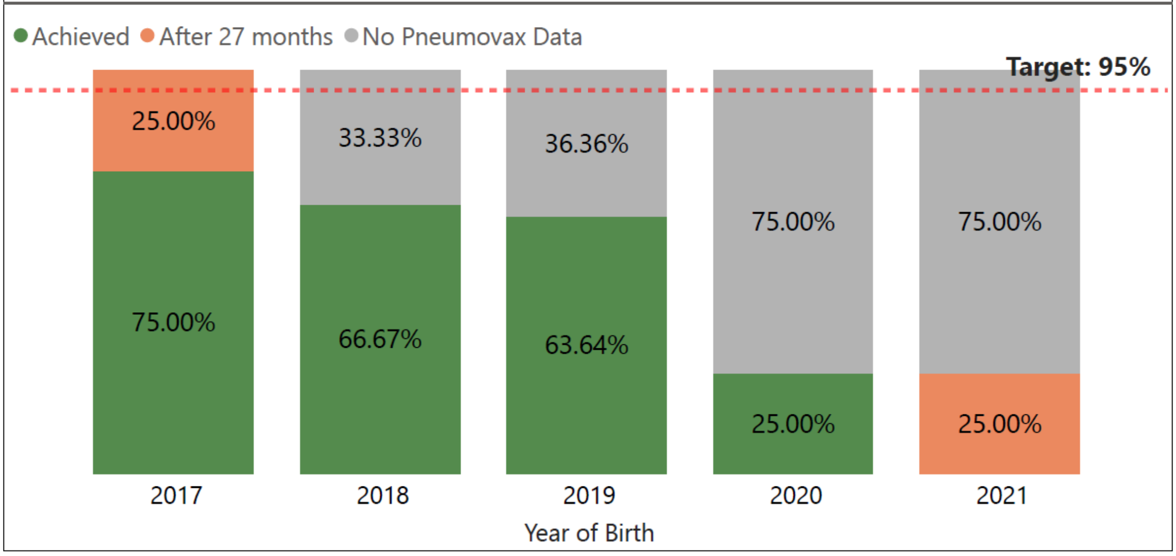
KPI 3 - 100% of patients with sickle cell disease are offered penicillin V (or alternative) by 3 months of age.



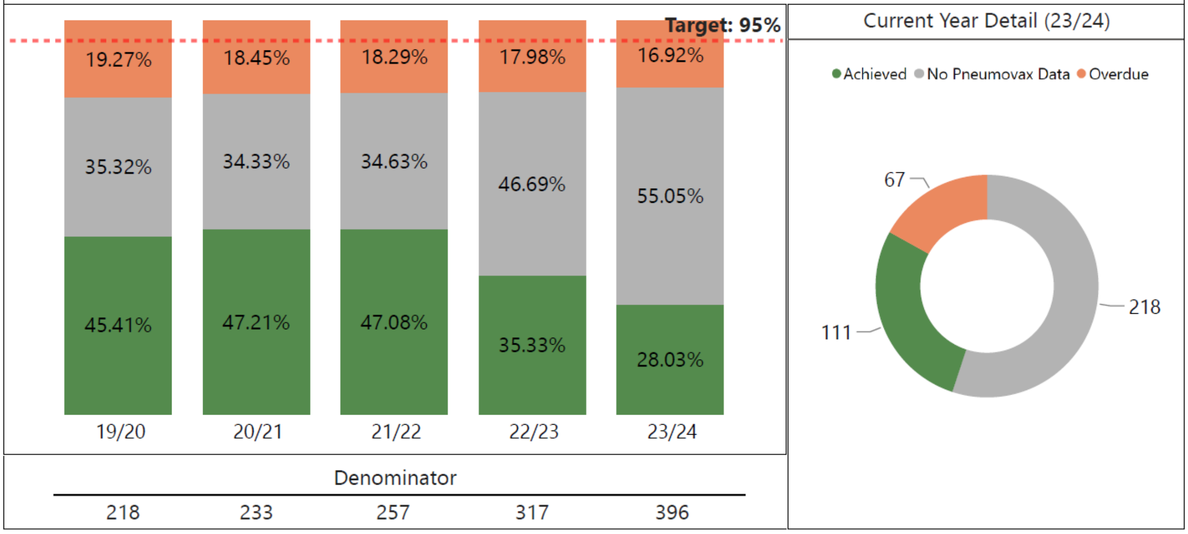
KPI 4 – 95% of patients should be given first Pneumovax (polycaccharide antigen) by 27 months and 5 yearly thereafter.

Access to data around vaccinations is recognised as challenging. At the strategic planning event it was agreed that further work was required to better understand they systems used for recording vaccinations and explore if there was a more effective way to access data and information. This is part of the workplan for 2024/25. It should be noted that the clinicians do not believe that a significant number of patients are missing out on vaccines, but that the data is not being captured.

KPI 4.1 - Percentage of patients given first Pneumovax (polysaccharide antigen) by 27 months of age.

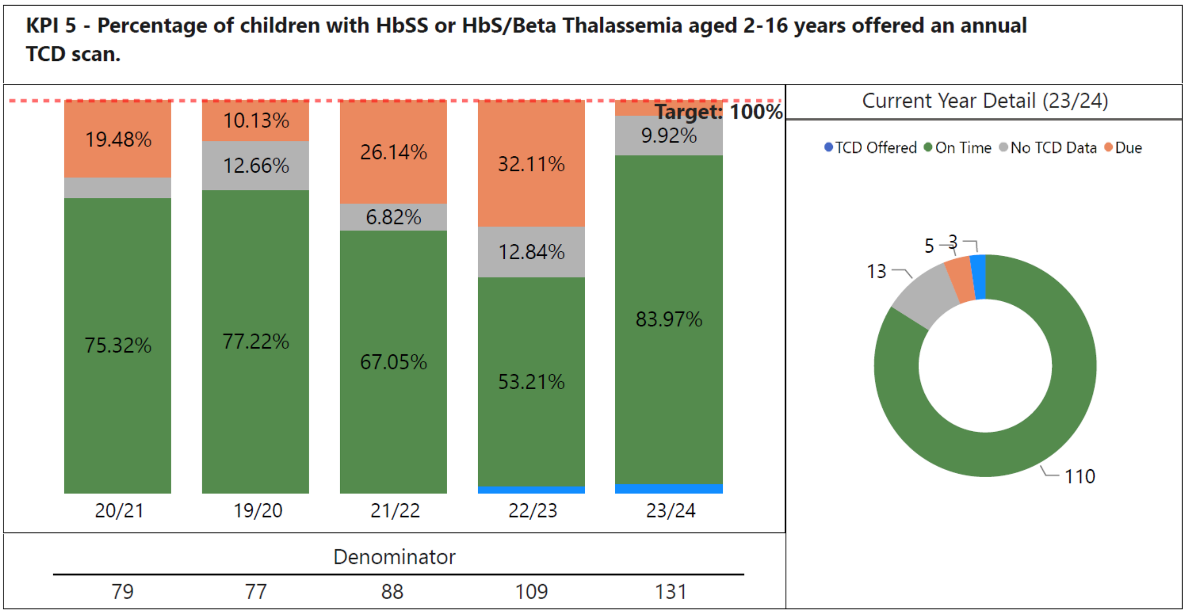


KPI 4.2 - Percentage of patients aged 2 years and over given Pneumovax (polysaccharide antigen) within the last 5 years.

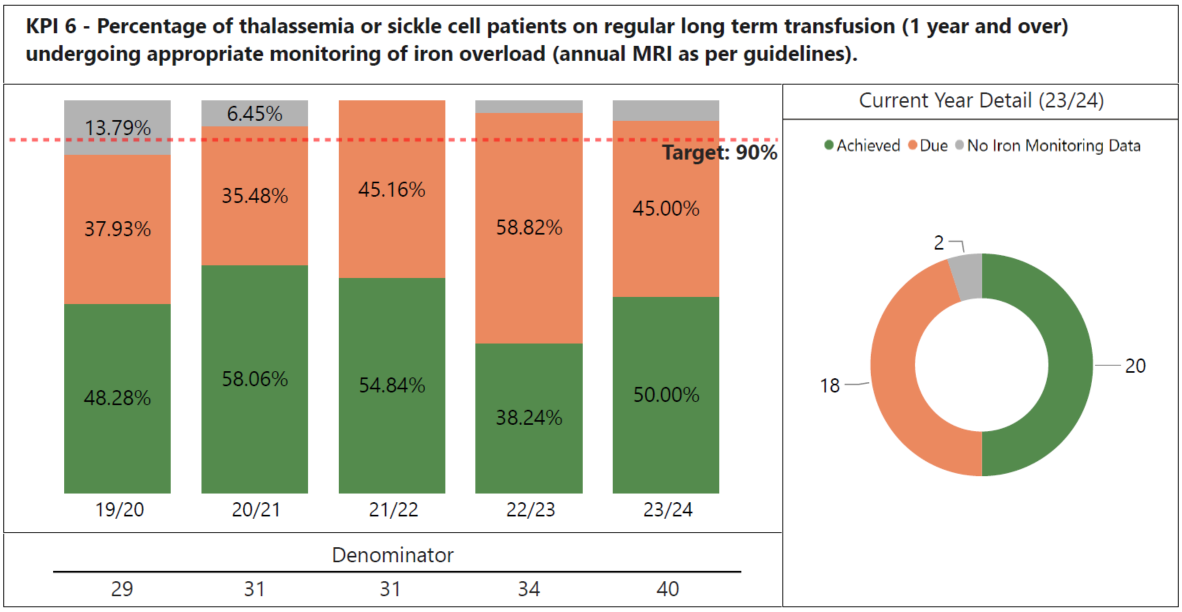


KPI 5 – 100% of children with HbSS or HbS/Beta thalassemia aged 2-16 years offered an annual TCD scan.

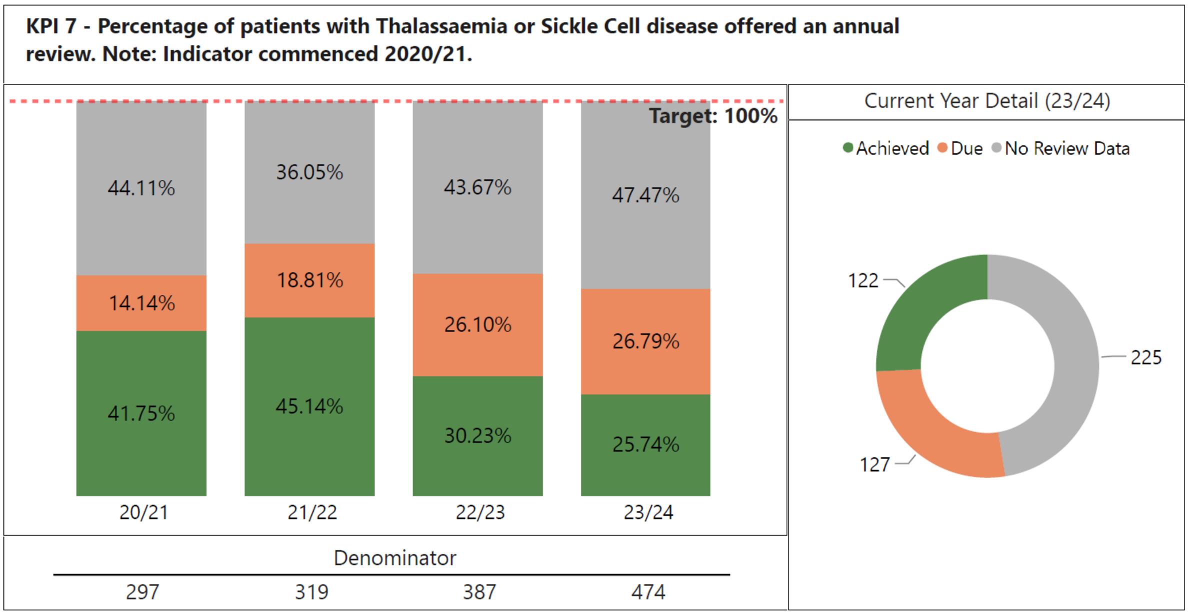
The TCD service have worked hard to catch up on annual scans following the pandemic and this is reflected in the significant increase in TCDs this year. Some additional funding was made available from NSD to increase the number of sessions which has also helped.


KPI 5 - Percentage of children with HbSS or HbS/Beta Thalassemia aged 2-16 years offered an annual TCD scan.

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

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



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 – 2024/25

Over the course of 2024/25 the programme of work will include:

- Developing best practice guidance for transition from paediatric to adults services
- Holding a workshop to engage with patients and families to understand what matters to them (delayed from 2023/24)
- Engaging with emergency departments to explore standards and improvements
- Developing education and pathways to support access to manual and automated red cell exchange
- Investigating whether information flows between specialist services and vaccination services can be improved
- Reviewing information provision to ensure that suitable information on PenV and folic acid is available for GPs and pharmacists
- Working with the TCD service to explore opportunities to increase accessibility of TCD and Ferriscan for children
- Developing referral and follow up pathways for transplant
- Providing advice on developing and delivering a service for patients in Scotland eligible for gene therapy to inform planning
- Exploring the level of need for psychology input for patients with haemoglobinopathies in Scotland and the options for delivery

Finance

The network spent £898 of their £5,000 budget, £800 on the strategy event and £98 on printing of leaflets for schools.