National Haemoglobinopathy Registry Report 2013/14

September 2014

Contents

Abbreviations

CCG	Clinical Commissioning Group
CRG	Clinical Reference Group
LAT	Local Area Team
MDSAS	Medical Data Solutions and Services
NHR	National Haemoglobinopathy Registry
NICE	National Institute for Health and Care Excellence
TCD	Transcranial Doppler

Complied by

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CHAPTER 1

NHR Overview

Dr Rob Hollingsworth

1.1 Success

With over nine thousand patient registrations the National Haemoglobinopathy Registry (NHR) is, in terms of the number of Haemoglobinopathy patients registered, the most successful registry of its type ever in the UK. This success is due to the considerable efforts of all the stakeholders involved, including patient societies (Sickle Cell Society and UK Thalassaemia Society), clinicians and NHS England. Treatment centres deserve particular credit having worked hard to consent patients and populate the NHR, initially in many cases without the provision of any additional resources to do so.

1.2 Registrations

However, although significant progress has been made in the registration of patients there is still much work to be done to ensure we have as complete and useful a registry as possible. With respect to this, this year sees the launch of the new annual review system for the NHR. It is essential that we collect annual data on patients to maintain the accuracy of patient data and to gather much needed outcome data with which to assess the effectiveness of treatment approaches. More complete reporting of serious events via the NHR is also a key area which can provide extremely valuable data and is simple and quick for treatment centres to report. Also launched this year is a new card for patients which treatment centres can produce directly from the NHR and which contains key information of importance to patients in need of urgent care.

Looking to the future, as the registry becomes more complete it is envisaged that the NHR will become the central data collection platform for haemoglobinopathies. This will have many benefits including the simplification of data collection exercises and a significant reduction in duplicated effort. The data from the NHR is already being used in significant efforts to improve resources for haemoglobinopathies.

1.3 Annual Report

This annual report is the first ever produced by the NHR. It provides an overview of data collected thus far, together with short reports from NHR stakeholders, some of which utilise the data from the NHR. Future NHR reports will expand on the information contained in this report and look to provide more detailed analytical data.

To conclude, going forward the NHR is the most important tool we have to improve the management of patient services, plan for the future, facilitate treatment research and ultimately improve patient care.

CHAPTER 2

Clinical Reference Group & the NHR

Dr Phil Darbyshire

2.1 Clinical Reference Group & The NHR

This year saw a reinvention of the Haemoglobinopathy clinical reference group (CRG), perhaps unwisely I volunteered to chair this group for another two years! This group essentially advises NHS England on specialist services for Haemoglobin disorders, it is responsible for drawing up policies and specifications to support cost effective and equitable commissioning of these disorders. Jon Currington describes in another article the quite dramatic changes that have taken place in the NHS landscape this last year and how they affect the commissioning of services. These changes should mean that we see improvements in access to care for our patients as well as improvements in the quality of care and improved outcomes for our patient group. One critical component of the new changes is the fact that patients are at the centre of the new structures , this hopefully will mean they are able to be closely involved in service design and provision.

This last year has seen a lot of activity attempting to incorporate the NHR as the key platform in collecting data on a national basis. It is increasingly evident that if we are to be successful in improving outcomes nationally then we must be collecting high quality data of value to all involved groups. Good data is vital for health professionals, we still have only a relatively poor overview of the natural history of Haemoglobin disorders in England, and the NHR should contribute to a better long term view. Clinicians, patients and commissioners need to know if any health intervention works, plus is it cost effective, again good long term data will provide this information. The CRG has recently been working with the NHR to ensure that the NHR registration and crucially the annual review fields are fit for purpose, they have been revised recently by Kate Ryan and others and now do have a close fit with key components of the Haemoglobinopathy specification, i.e. they fit in closely with what Trusts should be providing and they record the key outcomes.

Another key piece of data collection which is still being refined is the use of the NHR as a platform for individual patients to record, via a structured patient survey, their views on the services they are offered . It is hoped that this will provide a very different and independent view and will we hope be a valuable cross check on information being collected by other routes. An example might be pain control in Sickle cell disease. Centres are going to be asked to provide evidence of adherence to recent NICE guidelines on pain management, if it is found that a centre is saying it complies with guidelines 90% of the time but patients via the survey say there is only 20% compliance then clearly questions will need to be asked. We hope this survey will be a regular annual collection of data directly by patients; the aim is to begin early in 2015.

2.2 Adverse Events

One of the successes of the NHR has been the collection of information on serious events. The primary aim is educational, each Trust already has systems in place to learn from such events and the NHR system does not seek to replace such systems. We hope however that a wider view will show up lessons that might not be so obvious from individual cases. A new protocol has been drawn up in conjunction the Anne Yardumian at the UK Forum , this will be circulated widely and again we hope will go live within a few months.

2.3 Quality Dashboards

The final piece of the data 'jigsaw' is a quality dashboard. This is essentially an in year check to ensure that Trusts are meeting service specifications, e.g. are they meeting their targets with Transcranial Doppler screening? This dashboard is not run by the NHR, however there is a dialogue going on to ensure data is not entered in two separate areas.

CHAPTER 3

Commissioning

Jon Currington

3.1 New Commissioning structure

On 1 April 2013, NHS England was established with an overarching role to ensure the NHS delivers better outcomes for patients within its available resources, and upholds and promotes the NHS Constitution. The introduction of new commissioning arrangements means that the majority of NHS services in England are now commissioned by more than 200 Clinical Commissioning Groups (CCG). However, NHS England itself also has a substantial and legal responsibility for commissioning certain "prescribed" services. These prescribed services include armed forces healthcare; primary health (including dentistry); offender healthcare and specialised services.

Specialised services are defined by four criteria: the number of individuals who require provision of service; the cost of providing the service or facility; the number of persons able to provide the service or facility and the financial implications for CCGs if they were required to commission the service or facility themselves.

Haemoglobinopathy care, delivered by specialist acute centres either at the centre or as an outreach service, is one of the prescribed specialised services directly commissioned by NHS England.

It is the ambition of NHS England to achieve equity and excellence in the provision of specialised care and treatment which is patient centred and outcome based; fair and consistent throughout the country; ensures that patients have equal access to services regardless of their location, and improves productivity and efficiency.

This ambition is accelerated by: the changes to commissioning arrangements allowing a single national commissioning structure and process to be established enabling a focus on engaging with, and better understanding the needs of patients and carers; designing a clinically led system developing national strategies for services which are then locally delivered, and the opportunity to manage new innovations and the introduction of technologies in a systematic way.

3.2 Clinical Reference Groups

To provide NHS England with expert advice on its commissioning decisions, 75 Clinical Reference Groups (CRG) have been established. Membership of each of the CRGs includes clinicians from across England, patient and public representatives, and organisations with a role in quality or training, as well as commissioners. In addition, any interested parties or individuals are encouraged to register as a stakeholder in the CRGs. The Haemoglobinopathy CRG is chaired by Dr Phil Darbyshire supported by me as accountable commissioner.

The CRGs are organised into four programs of Care; the Haemoglobinopathy CRG is part of the Cancer and Blood Program of Care which is chaired by Sean Duffy, National Clinical Director for Cancer.

As part of their advisory role to NHS England, the CRGs are responsible for producing commissioning products such as commissioning policies and service specifications. The service specification for Specialised Haemoglobinopathy, which has been subject to formal consultation, was implemented as part of NHS England contracts from October 2013. This specification describes what services should look like for patients and what they should expect. Implementation and monitoring of compliance to the specification is the responsibility of NHS England's 10 area teams which have a specific responsibility for specialised services. Where a hospital does not meet the specification, a time limited plan (or derogation) to address any shortfalls will be agreed between the area team and hospital.

In its first year as a direct commissioner of specialised services, NHS England's CRGs have produced more than 120 service specifications for implementation by area teams to achieve a nationally consistent approach to commissioning, differing regional and local approaches to contracting with providers are being brought together. Resolving different sets of standards, and varying levels of access to all services, including Haemoglobinopathy, around the country is a challenge for now and the next year. The use of the National Haemoglobinopathy Register is a requirement of the specification and, if universally adopted and completed, would be a useful tool in meeting these challenges and improving services for patients

CHAPTER 4

Patient Societies

John James & Gabriel Theophanous

4.1 Sickle Cell Society

"WORKING TOGETHER FOR PATIENTS OF SICKLE CELL AND THALASSAEMIA"

The development of the National Haemoglobinopathies Register (NHR) is a welcome initiative offering an inclusive focus for all key stakeholders involved with Sickle Cell and Thalassaemia. It is encouraging to see the creation of a forum that engages Sickle Cell and Thalassaemia patients, their doctors and nurses, NHS managers and patient organisations, such as the Sickle Cell Society, to develop such a vital database that will help the planning, development and delivery of services to patients with sickle cell disease or Thalassaemia.

We have been particularly impressed by the NHR facility that has enabled doctors, nurses and managers to learn from adverse event reports. We are also encouraged that the NHS Clinical Reference Group for Sickle Cell and Thalassaemia has taken an interest in the NHR. We are pleased that the NHR is also keen to engage with corporate bodies on developments in biotechnology with reference to the Haemoglobinopathies. This is key, for example, with the development of technologies that allow for automated red cell exchange to be available throughout the UK, providing universal access to patients.

We, at the Society, fully support the development of the NHR. We hope the NHR will become the single and most comprehensive data source for the NHS to work with stakeholders to align advocacy, strategy and programs to improve services to individuals with Sickle Cell and Thalassaemia. This will help us to ensure that the NHS continues to improve and respond to the needs of Sickle Cell and Thalassaemia patients.

We look forward to working within the NHR framework, encouraging the take-up by Sickle Cell patients and their families, our stakeholder partners and all contributing to advancing improvements in blood services.

John James
CEO
Sickle Cell Society

4.2 Patients Group Feedback

"ENCOURAGING A SUSTAINABLE PARTNERSHIP ON BLOOD DEVELOPMENTS IN THE NHS"

The development of the National Haemoglobinopathies Register is a welcome initiative as it seems to be offering already an inclusive focus for all the key stakeholders. It is encouraging to note the emergence of a forum that engages sickle-cell and thalassaemia patients, their doctors and nurses, NHS managers and other key players all working together to develop such a vital database that would help the planning , development and delivery of responsive services to patients with sickle cell disease or thalassaemia.

On a note of substance, we have been particularly impressed by the facility that has enabled the doctors, nurses and managers to learn from adverse events reports. We are also encouraged that the nascent NHS Clinical Reference Group for sickle-cell and thalassaemia has taken keen interest in the NHR. And that the NHR is also keen to engage with corporate bodies, so as to inform developments in biotechnology with reference to the Haemoglobinopathies, for example technologies that allow for automated red cell exchange to be available throughout the UK providing universal access to patients.

We, at the Society, are hopeful that these developments will continue to clarify, in a single resource, the NHS vision for blood genetic disorders, so that together we can better align our advocacy strategy and programs to ensure that the NHS continues to improve and respond to the needs of Sickle Cell and thalassaemia patients. Together, therefore, we look forward to working within the NHR framework, encouraging the take-up by sickle-cell patients and their families, our corporate partners and all contributing to advancing improvements in blood services.

4.3 UK Thalassaemia Society

The UK Thalassaemia Society welcomes the annual report of the National Haemoglobinopathy Registry; and I am delighted to write a few words in support of the NHR. At the UK Thalassaemia Society we are often contacted by researchers and medical students who want information about thalassaemia in the UK; and the first thing they usually ask is "How many people in the UK have thalassaemia?" It always comes as a surprise to them that there is no answer to that question. As the recent peer reviews of Haemoglobinopathy services for children and adults have clearly shown, thalassaemia exists mainly in certain parts of the UK; and these are not necessarily the same areas which have a high prevalence of sickle cell patients. The peer reviews have also made it glaringly obvious that the quality of services for the treatment of thalassaemia show huge variations in quality depending on where the patients happens to be treated. In addition, we are aware that there are patients who remain unknown to the networks, being treated as lone outliers with no access to specialist services and expertise.

Unless we know exactly how many thalassaemia patients there are and just as importantly, where they are, we cannot work to bring the quality of services up to a consistent standard everywhere. The NHR provides the simplest and most obvious solution to these questions. The UK Thalassaemia Society believes that it should be a requirement for every Haemoglobinopathy patient to be entered onto the Registry, in every hospital. Only when we can track every patient, their treatment, annual reviews and eventual outcomes will we have the information to provide the correct and consistent treatment for all patients, regardless of where they are.

We hope that the changes in the NHS will result in more resources for our hard-pressed services to enable them all to enter their patients onto the NHR. The Peer review program 2012-13 overview Report (adult services) states "The NHR provides a means of monitoring the number of affected adults in a clinical centre, for adverse event reporting and for demonstrating compliance with key standards of clinical care. A lack of data administration support was a common barrier to data collection." (pp 4-5). A relatively small investment now in better administrative support for clinical staff could result in huge savings in future years if patient care is standardised and outcomes improved. The NHR gives us the opportunity to obtain the information we need to achieve this result; and it is an opportunity that must not be wasted.

Gabriel Theophanous

President, UK Thalassaemia Society

CHAPTER 5

Sickle Cell Reports

Mark Foster

5.1 Reports Overview

This chapter aims to provide a quick overview into Sickle Cell patient numbers for England. All data is for patients registered on to the NHR by 31st March 2014. For future Annual Reports it is hoped to utilise data generated from the Annual Reviews. This will be achievable with the further integration of Annual Reviews into the NHR and improved completion rate of Annual Reviews by centres.

As of March 2014 49 centres have registered Sickle Cell patients on to the NHR with a total of 7338 patients. Around 300 patients are registered at more than one centre. As the NHR is a consented registry we only have records of patients who have offered their consent. Although the rate of patients that refuse consent is low it is vital that we record non-consenting patients numbers at each centre so we can maintain accurate patient numbers.

5.1 Commissioning Changes

Following changes to the commissioning structure in England, MDSAS developed new map templates allowing for the reporting of patient numbers by Commissioning Hub and local area team (LAT). Figure 5.4.1 shows number of Sickle Cell patients by commissioning hub.

A full list of Sickle Cell reports is provided in the table below

Figure 5.1 Centres with highest numbers of registered Sickle Cell patients

Centre	No. Patients
Guy's and St Thomas	885
Kings College Hospital	801
North Middlesex	499
UCLH and Whittington hospital	498
Queens Hospital (BHR)	361
Homerton Hospital	319
North West London Hospitals NHS Trust (Central Middlesex Hospital)	306
Birmingham Children's	292
Queen Elizabeth Hospital, Woolwich	256
Manchester Royal Infirmary	229
Manchester Children's Hospital	223
Imperial College Healthcare NHS Trust (Hammersmith Hospital)	194
St Georges Healthcare NHS Trust, London	188
Birmingham - City Hospital	178
Croydon Health Services NHS Trust	170
Imperial College Healthcare Trust (St Mary's)	168
University Hospitals Leicester	127
Lewisham Healthcare NHS Trust	126
Royal London	123
Nottingham University Hospitals	120

Figure 5.2 Sickle Cell diagnosis type chart

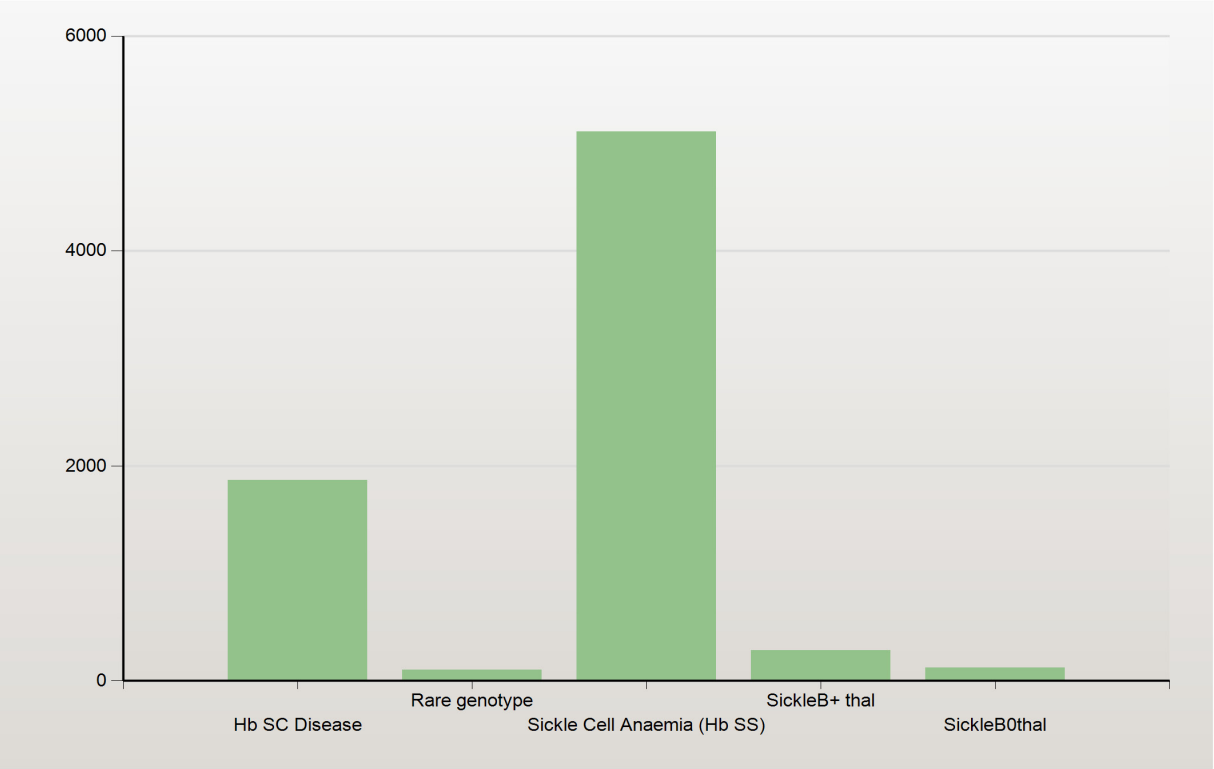


Figure 5.3 Sickle Cell patients ethnicity table

Ethnicity	No. Patients
African	4434
Caribbean	1477
Any other Black background	600
Not Stated	529
White and Black African	96
Any other ethnic group	89
White and Black Caribbean	62
Any other mixed background	55
Indian	46
Any other Asian background	43
Any other White background	22
White - British	16
Pakistani	11
Bangladeshi	< 10
White and Asian	< 10
White - Irish	< 10

Figure 5.4.1 Map of Sickle Cell patients by commissioning hub

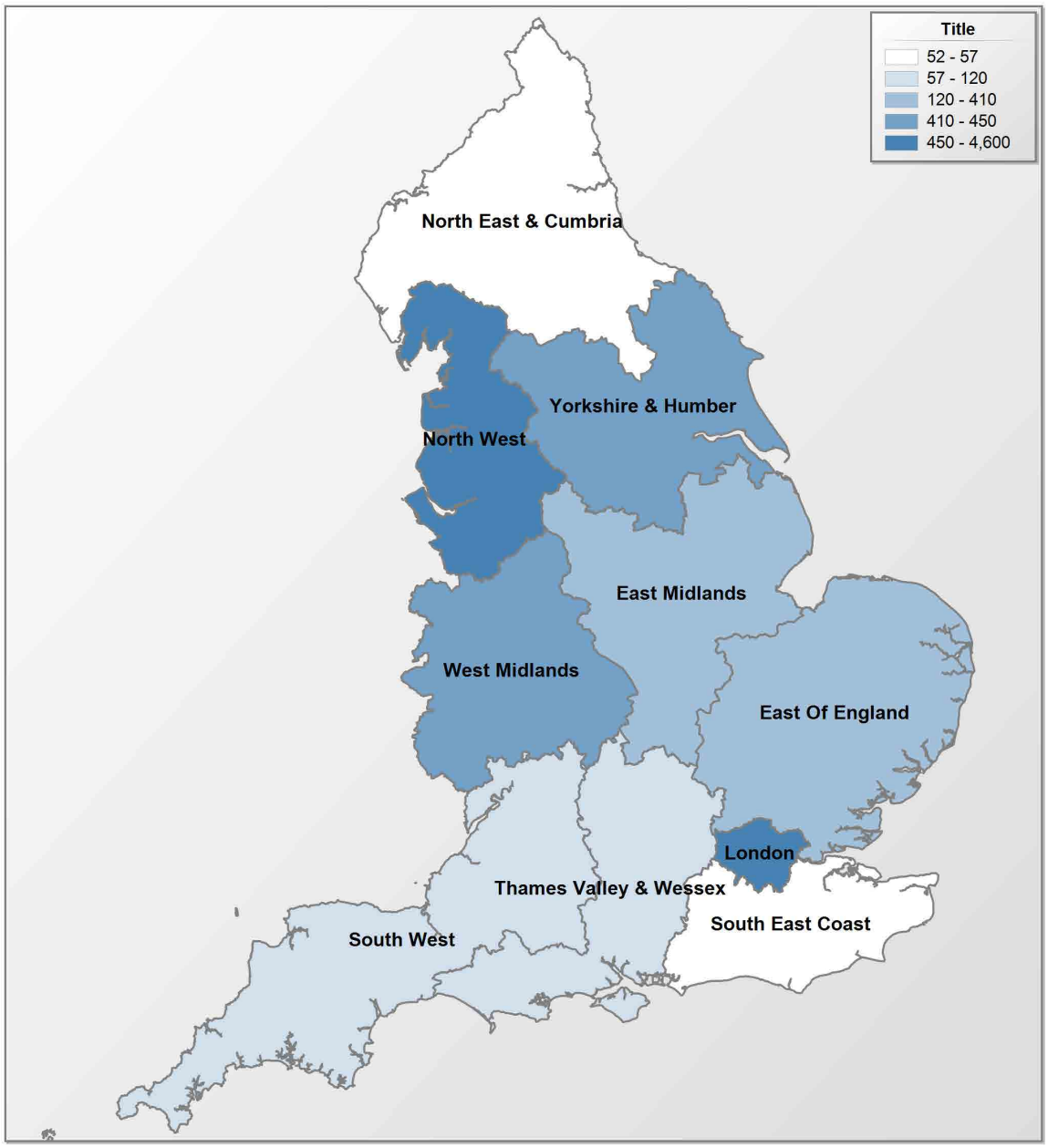


Figure 5.4.2 Table of Sickle Cell patients by commissioning hub

Region	No. Patients	Region	No. Patients
London	4558	East Of England	146
North West	489	Thames Valley & Wessex	113
Yorkshire & Humber	419	South West	63
West Midlands	412	North East & Cumbria	52
East Midlands	380	South East Coast	52

Figure 5.5 Sickle Cell Patients by Current Age

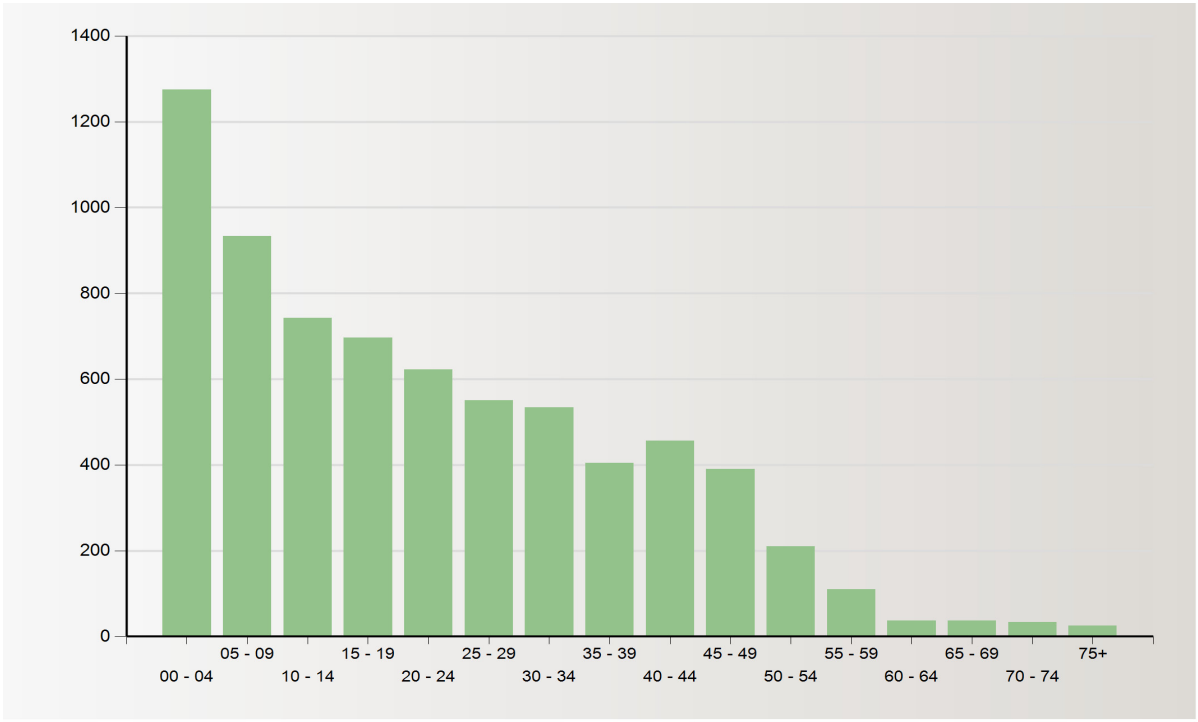


Figure 6.6 Sickle Cell therapy type chart

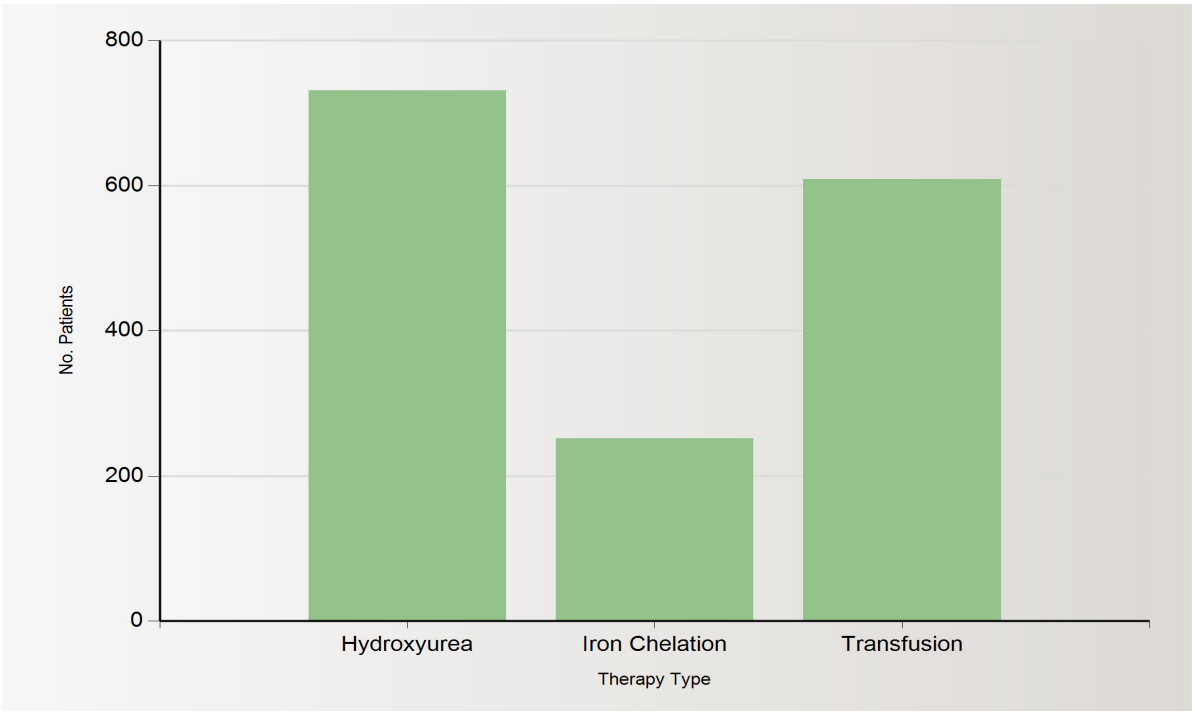


Figure 5.7 Sickle Cell iron chelation type chart

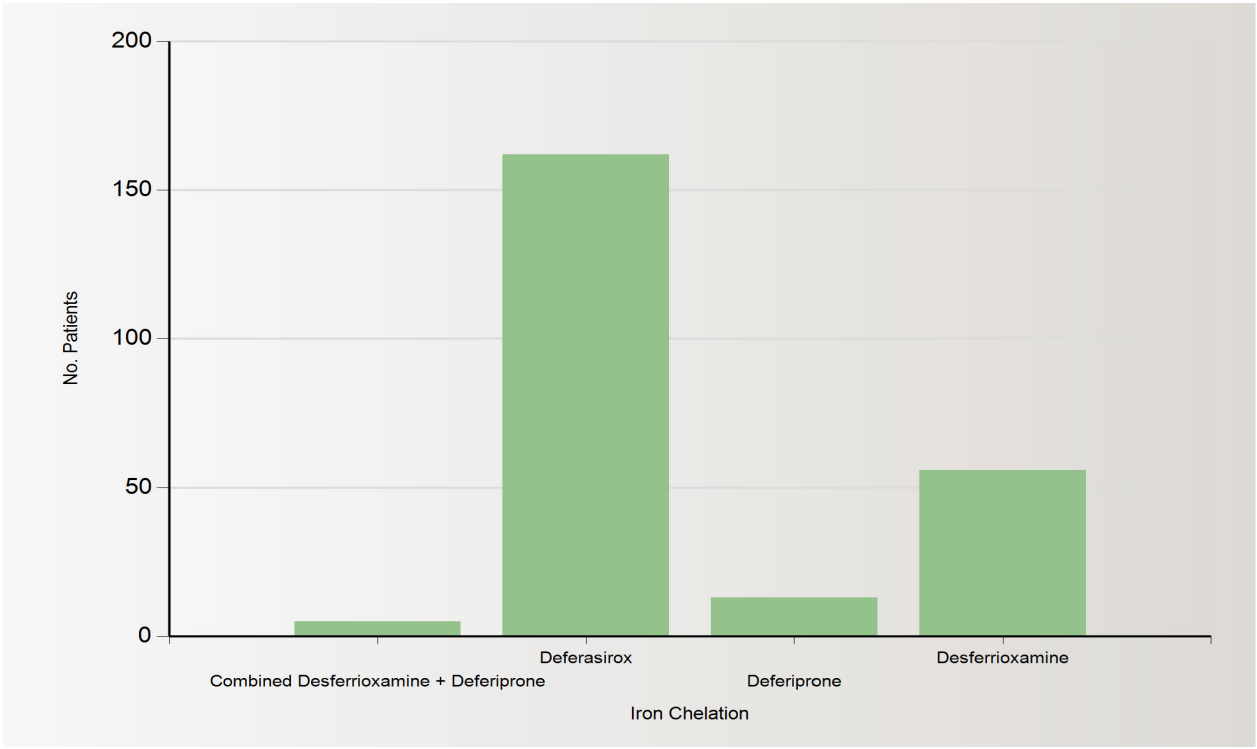


Figure 5.8.1 Map of patients with TCD Monitoring by Commissioning Hub

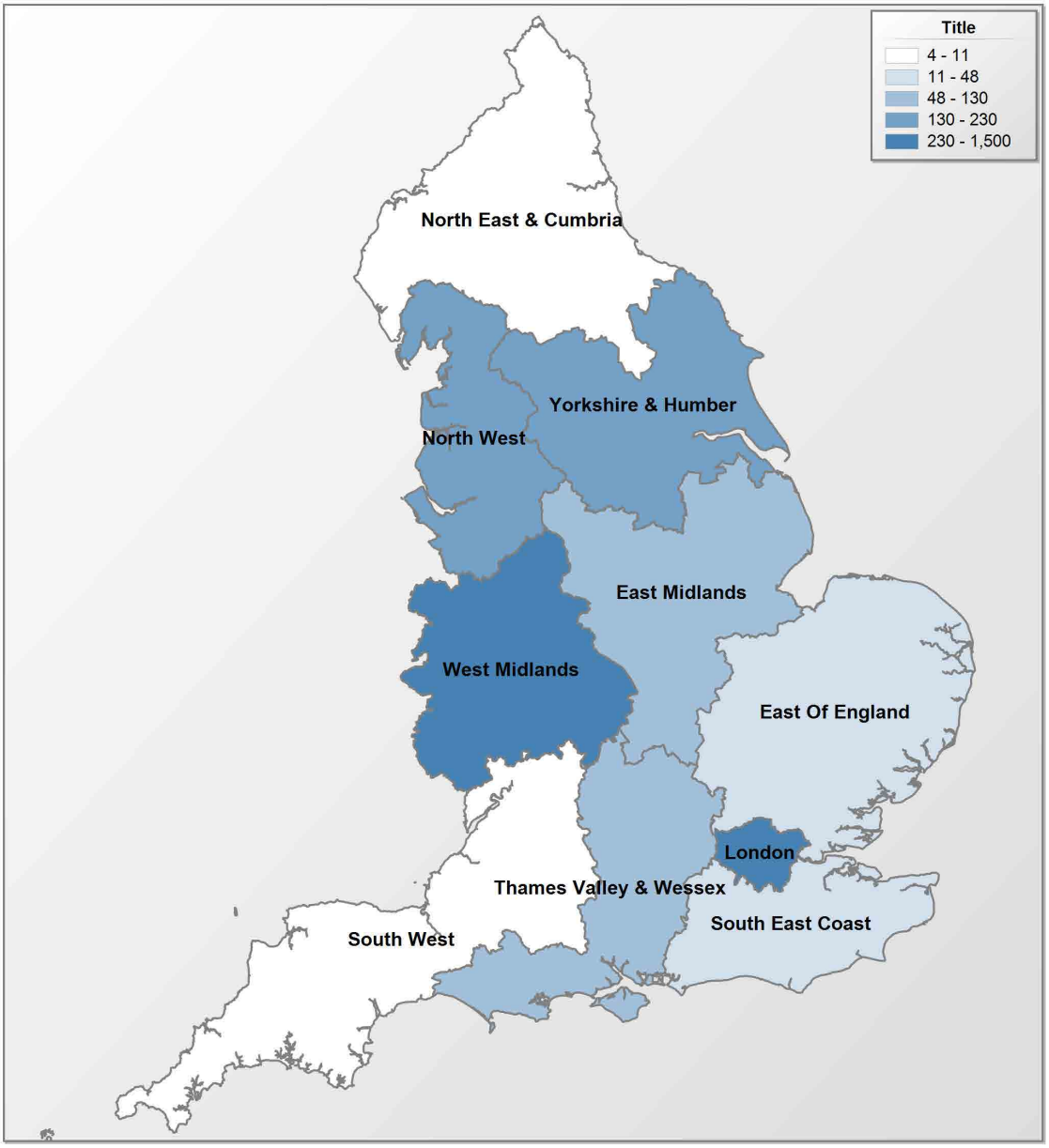


Figure 5.8.2 Table of patients with TCD Monitoring by Commissioning Hub

Region	No. Patients	Region	No. Patients
London	4558	Thames Valley & Wessex	146
West Midlands	489	East Of England	113
North West	419	South East Coast	63
Yorkshire & Humber	412	North East & Cumbria	52
East Midlands	380		

CHAPTER 6

Thalassaemia Reports

Mark Foster

6.1 Reports Overview

As with the Sickle Cell chapter all data presented for Thalassaemia is for patients registered onto the NHR by 31st March 2014. AS the rate of Thalassaemia patients registered on the NHR is fairly high, the reports presented here provide an excellent overview on the Thalassaemia population in England. A full list of Thalassaemia reports is provided in the table below. As of March 2014, 39 centres have registered Thalassaemia patients on the NHR with a total of 906 patients. 50 patients are registered at more than 1 centre.

Figure 6.1 Centres with highest numbers of registered thalassaemia patients

Centre	No. Patients
UCLH and Whittington Hospital	232
Birmingham Children's	133
Manchester Children's Hospital	52
Manchester Royal Infirmary	50
Imperial College Healthcare Trust (St Mary's)	43
Birmingham - City Hospital	38
North Middlesex	38
Bradford	37
University Hospitals Leicester	29
St James's University Hospital, Leeds	26
Kings College Hospital	23
Queens Hospital (BHR)	23
Luton and Dunstable Hospital	20
Coventry University Hospital	19
Imperial College Healthcare NHS Trust (Hammersmith Hospital)	14
North West London Hospitals NHS Trust (Central Middlesex Hospital)	14
Sheffield Children's Hospital	13
University Hospitals Bristol NHS Trust	13
Nottingham University Hospitals	10
St Georges Healthcare NHS Trust, London	10

Figure 6.2 Thalassaemia diagnosis type

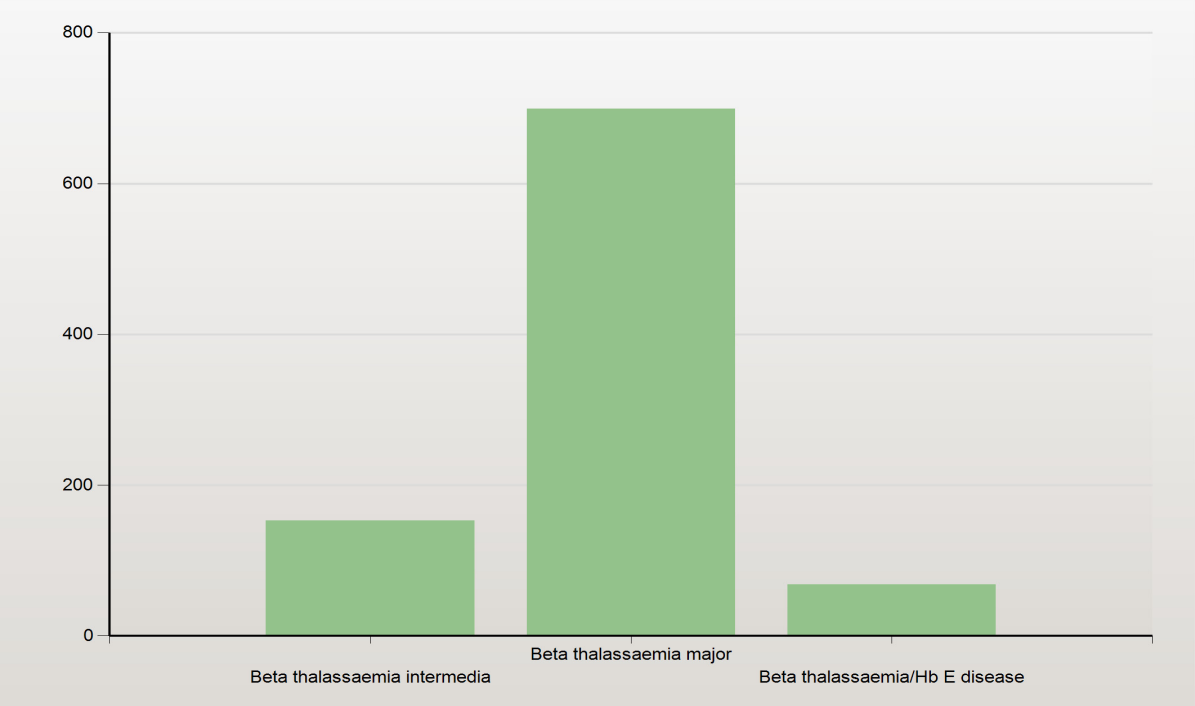


Figure 6.3 Thalassaemia patients ethnicity table

Ethnicity	No. Patients
Pakistani	353
Indian	126
Any other ethnic group	92
Any other White background	92
Any other Asian background	79
Not Stated	53
Bangladeshi	43
White - British	26
Chinese	15
African	11
Caribbean	11
Any other mixed background	< 10
White and Asian	< 10
Any other Black background	< 10
White and Black Caribbean	< 10

Figure 6.4.1 Map Thalassaemia patients by commissioning hub

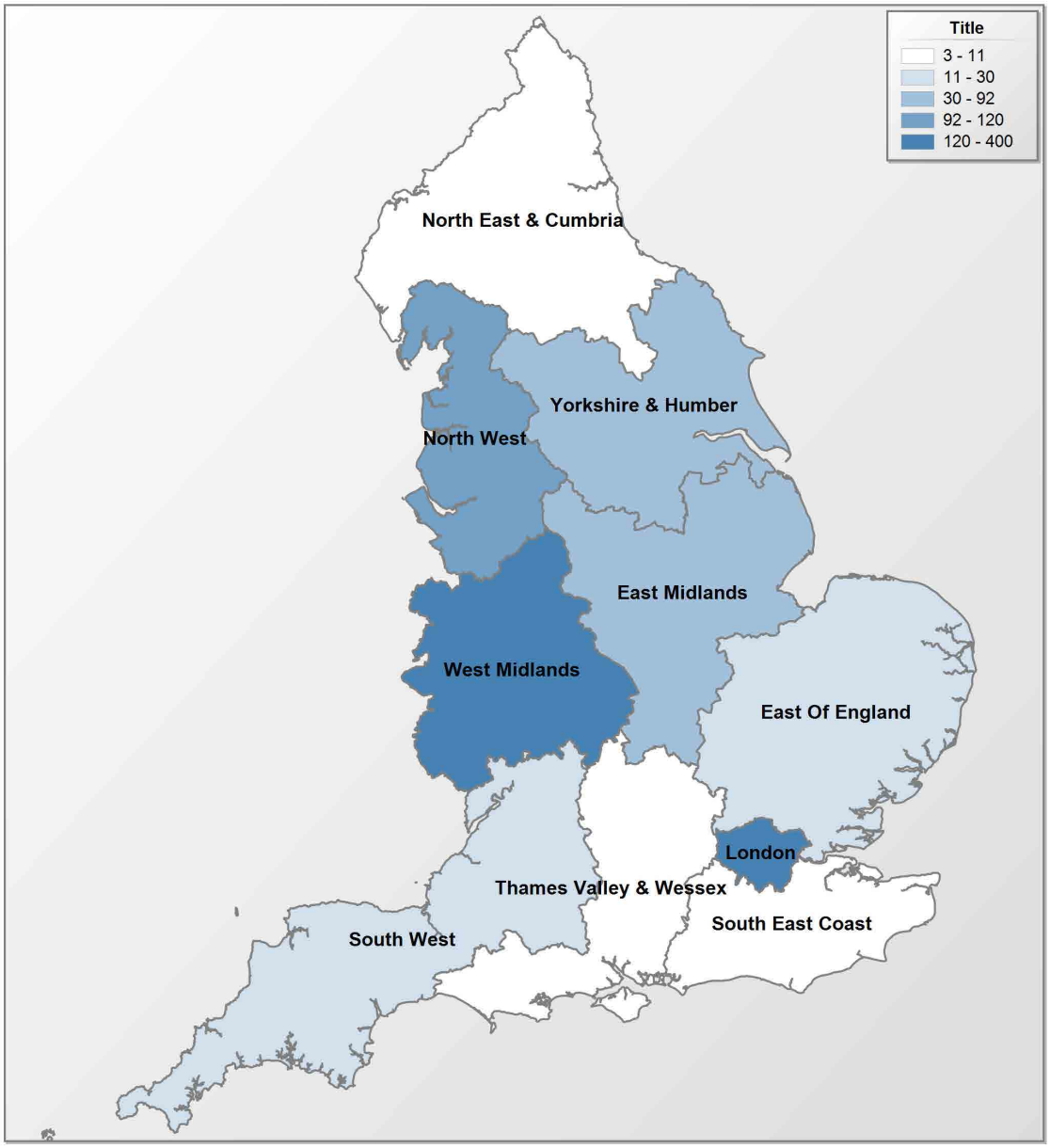


Figure 6.4.2 Table Thalassaemia patients by commissioning hub

Region	No. Patients	Region	No. Patients
London	4558	East Of England	146
North West	489	Thames Valley & Wessex	113
Yorkshire & Humber	419	South West	63
West Midlands	412	North East & Cumbria	52
East Midlands	380	South East Coast	52

Figure 6.5 Thalassaemia patients by age group

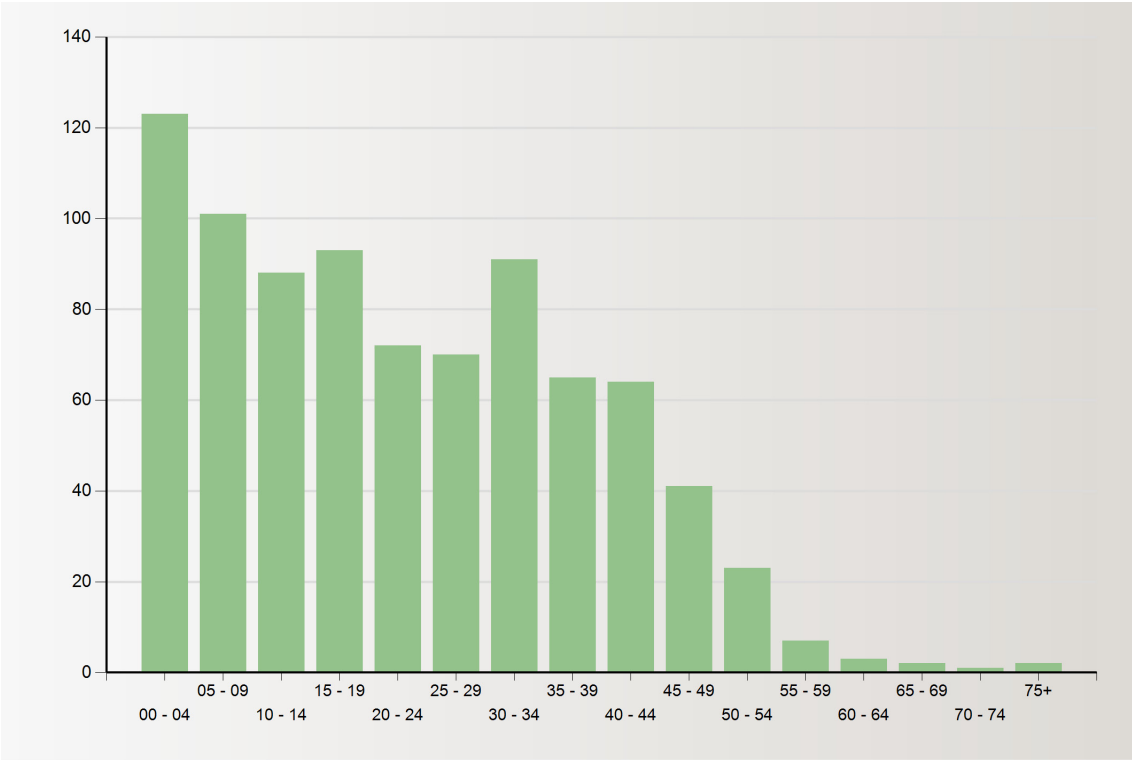
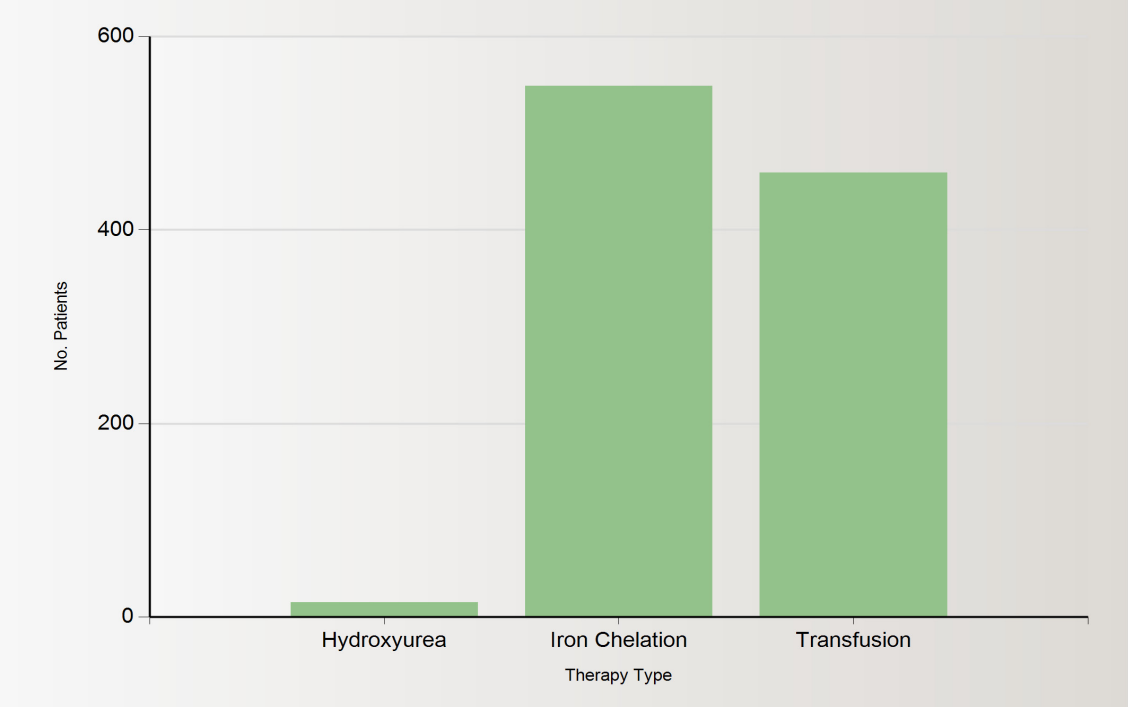


Figure 6.6 Thalassaemia therapy type chart



CHAPTER 7

Transfusion Data

Sara Trompeter

7.1 Introduction

There are limited data on the NHR with regards to transfusion. In the initial dataset, when registering patients, the following transfusion relevant data is requested:

- Therapy (transfusion): if this is selected then there is a follow up question regarding estimated lifetime transfusion exposure (0-10, >20-50, >50 units)
- Regular transfusion: (tick for yes)
- Year of first transfusion

In the annual review there is data collected on:

- Transfusion in the study period
- Therapies started or continued: (automated exchange, manual exchange, and top up).

For this report we will only analyse the data from the initial dataset as the annual review was only completed for a very small minority of patients and 50% of these patients came from one hospital, making external validity poor.

7.2 Sickle Cell - Baseline data

Of the 7488 patients with sickle cell disease 606 have their transfusion history recorded in their baseline data. We have grouped those with unknown and no transfusion data together. The question asked is 'what is the lifetime overall transfusion exposure?' This is divided into three groups; 0-20 units, >20-50 units, >50 units. From the data collected, there are 197, 140 & 256 in these three groups respectively. By breaking this down into the different sickle genotypes, we can see that of the 606 patients with SCD and Transfusion Data:

- HbSS 87%
- HbSC 8%
- HbSβ+ thal 2%
- HbSβ0 thal 2%
- Rare * 1%

However, note from the table below, only a very small proportion of patients in each group had their transfusion data recorded at all. Further to this the likelihood of having transfusion data recorded seemed to correlate with transfusion exposure, suggesting that the data may be skewed in favour of patients getting transfusions. This is not an impossible inference as they attended hospital more frequently and are therefore more likely to be consented for NHR. Thus the likelihood of having data included is in theory increased.

Table 1 Proportion of difference sickle genotypes with baseline transfusion data recorded

Diagnosis	Transfusion Data	No Data	Total
HbSS	528 (10.3%)	4580	5108
HbSC	52 (2.9%)	1819	1871
HbSβ0 thal	13 (4.6%)	270	283
HbSβ+ thal	13 (8.1%)	113	123
Rare Genotypes*	3 (2.9%)	100	103
Total	**606 (8.9%)**	**6882**	**7488**

* Hb S/D Punjab, Hb S/E, Hb S/HPFH, Hb S/OArab & HbS/Lepore make up the rare sickle genotypes.

7.3 Age of first transfusion

A further question asked is 'what is the year of first transfusion?' To better understand this in real terms it is helpful to know the age at first transfusion. For those with sickle cell disease the age at first transfusion was filled out for 396 of 7488 patients. The group data for Sickle Cell shows that a third of patients are receiving their first transfusion before school age.

By breaking this down further into the different Sickle Cell genotypes, one can see that patients with HbSS and HbSβ0 thal are most likely to have their first transfusion in childhood whereas those with HbSC are most likely to have their first transfusion in adulthood. Patients with HbSβ+ thal were more likely to have their transfusion in adulthood. However, this effect is less marked than for patients with HbSC; 40% of patients have their first transfusion in childhood. Note once again that the statistical validity of this is poor due to the limited data from the transfusion fields. Note also that as HbSS contributes most to the transfusion episodes, the overall data is skewed to reflect HbSS.

7.4 Changing practice

The data (see figures 1 and 2) suggests that transfusion practice is changing. It looks as if larger proportions of patients have received their first transfusions at younger ages as time has moved on. However, this data is biased. Those with severe disease phenotypes, and thus those more likely to have received transfusions at a younger age, may well not have survived into the older cohort. Therefore the older patient cohort may represent those that, at least early in life, had a mild disease phenotype.

Table 2: Age at which patients received their first transfusion

Age	Overall	HbSS	HbSC	HbSβ0 thal	HbSβ+ thal
<6	127	118 (34%)	2 (7%)	4 (40%)	3 (30%)
6-15	118	113 (33%)	1 (3%)	3 (30%)	1 (10%)
16-29	104	82 (24%)	16 (55%)	2 (20%)	4 (40%)
30-49	45	34 (10%)	8 (28%)	1 (10%)	2 (20%)
50+	2	0	2 (7%)	0	0
Total	**396**	**347**	**29**	**10**	**10**

Figure 1: Sickle patients currently aged 30-49: Age at first transfusion

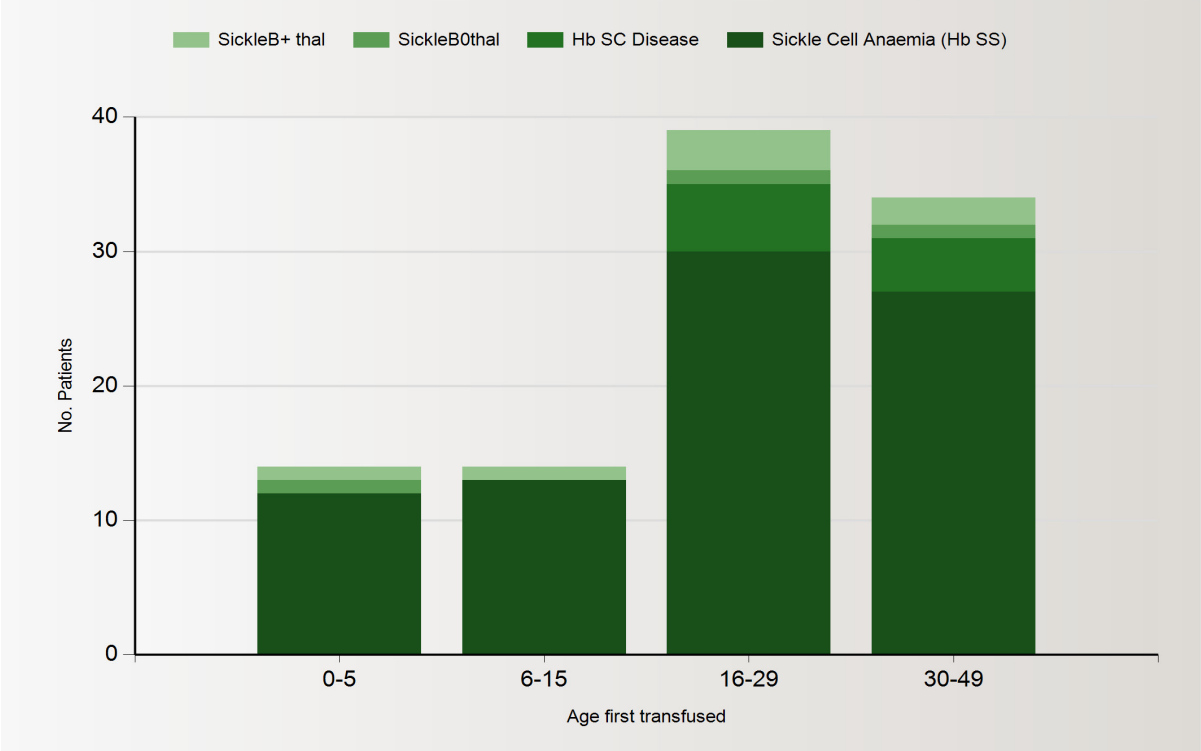
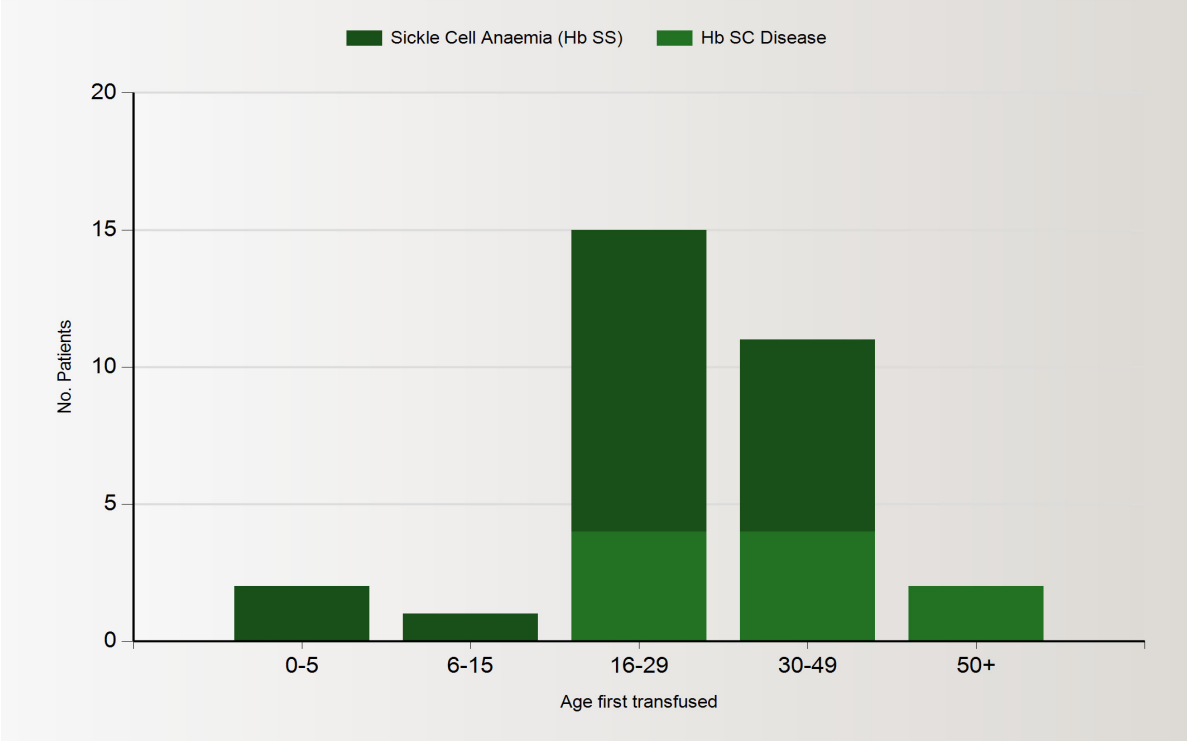


Figure 2: Sickle patients currently aged 50+: Age at first transfusion



7.5 Transfusion Exposure

Of the 606 patients with sickle cell disease who had transfusion data recorded, 593 had their transfusion exposure documented. Here we can see that 45% of patients who have their transfusion exposure recorded have had a lifetime excess of 50 units exposure. The HbSS patients skew the data here as they represent 87% of those with transfusion exposure recorded. However, they are the prominent patient genotype and even if they were represented at a slightly lower proportion in line with new born screening data or data from the NHR, this would not greatly affect this result. What would have affected the data was whether a patient being regularly transfused influenced the likelihood of them having their data recorded. The data suggests this may be the case. Of the 606 patients with sickle cell disease who had transfusion data recorded, 332 had, whether they were on regular transfusions or not, data recorded and 55% percent of patients are on regular transfusions. This far exceeds the data quoted elsewhere (10-20%). However, much of that data is also subjected to bias and inaccuracies and it may be that the number lies somewhere in between. It is only with a more complete data set that one can truly understand the data properly.

Table 3: Transfusion exposure and Sickle Genotype

Units	Overall	HbSS	HbSC	HbSβ0 thal	HbSβ+ thal	Rare
0-19	197	155 (30%)	33 (63%)	3 (20%)	6 (46%)	
20-50	140	119 (23%)	13 (25%)	2 (20%)	5 (39%)	1 (33%)
50+	256	241 (47%)	6 (12%)	5 (50%)	2 (15%)	2 (67%)
Total	**593**	**513**	**52**	**10**	**13**	**3**

Table 4: Patients on regular transfusion and Sickle Genotype

Regular Transfusion	Overall	HbSS	HbSC	HbSβ0 thal	HbSβ+ thal	Rare
Yes	332 (55%)	316 (60%)	6 (12%)	5 (50%)	2 (15%)	3 (100%)
No	274 (45%)	212 (40%)	46 (88%)	5 (50%)	11 (85%)	0
Total	**606**	**528**	**52**	**10**	**13**	**3**

7.6 Thalassaemia - Baseline Data

For thalassaemia the disease subtype options are beta thalassaemia major, beta thalassaemia intermedia and Beta thalassaemia/Hb E disease. This categorisation is not a helpful one. The latter group could be considered to have either of the former two groups' condition; traditionally thalassaemia major implies on a regular transfusion programme and thalassaemia intermedia implies not. A further complication is that patients over time can switch between these two groups as they become transfusion dependent with age. Here the most useful data would be from the annual review. The data collection for these conditions was much more complete with 50% of registered patients on the NHR having their transfusion data recorded, at least in part.

- Intermedia 6%
- Major 90%
- Hb E disease 4%

Of the 9210 patients with thalassaemia, 458 have their transfusion history recorded in their baseline data. The question asked is what is the lifetime overall transfusion exposure? This is divided into three groups; 0-20, >20-50, >50. From the data collected, there are 39, 61 and 356 in these three groups respectively. Thus the vast majority of patients where this data is recorded have been exposed to >50 units (78%).

7.7 Age of first transfusion

A further question that is asked is 'what is the year of first transfusion?' To better understand what this means it would be helpful to understand what is the age at first transfusion. For those with thalassaemia the age at first transfusion was filled out for 353 of 838 patients. Of those where the data is recorded, 92% had their first transfusion by 6 years of age and everyone had had a transfusion by 49 years of age. Many of the thalassaemia intermedia patients (68%) had also had a transfusion by school age. There was not enough data to see whether transfusion practice was changing in the intermedia and Beta thalassaemia/Hb E disease groups, however, in the thalassaemia major group there was no evidence of change in practice vis a vis age of first transfusion.

Breaking this down further into the different thalassaemia subtypes:

Year of first transfusion recorded:

- Major 76%
- Intermedia 16%
- Hb E disease 8%

Table 5: Patients with Thalassaemia who have their transfusion baseline data recorded

Diagnosis	Transfusion Data	No Data	Total
Beta thalassaemia major	409	290	699
Beta thalassaemia intermedia	33	120	153
Beta thalassaemia/Hb E disease	16	52	68
Total	**458**	**462**	**920**

Table 6: Age started treatment, Thalassaemia

Age	Overall	Major	Intermedia	HbEBeta
<6	324	299 (96%)	15 (68%)	10 (50%)
6-15	20	10 (3%)	3 (14%)	7 (35%)
16-29	5	2 (1%)	2 (9%)	1 (5%)
30-49	4	0	2 (9%)	2 (10%)
50+	0	0	0	0
Total	**353**	**311**	**29**	**10**

7.8 Transfusion Exposure

Of the 528 patients with thalassaemia who had transfusion data recorded, 456 had their transfusion exposure documented. Although the thalassaemia major patients have the highest transfusion exposure, it should be noted that the intermedia patients are not far behind. This may be due to three factors: firstly the data are small in number and may not be representable; secondly these patients were initially intermedia and now are major clinically; thirdly there is a cohort of patients receiving multiple ad hoc transfusions such that they have a considerable overall transfusion exposure. The latter would be concerning as it is often these patients who do not have their iron levels and chelation treatment subjected to as much scrutiny as those on regular transfusion programmes. In fact, looking at the data in table 7, 80% of the thalassaemia intermedia patients are being regularly transfused which would mean that these are actually thalassaemia major patients by most people's definition. Of the 438 patients with thalassaemia who had transfusion data recorded, 377 had, whether they were on regular transfusions or not, data recorded. This relates mostly to a couple of centres with large Thalassaemic populations.

Table 7: Transfusion Exposure, age started treatment, Thalassaemia

Units	Overall	Major	Intermedia	HbEBeta
0-19	39	32 (8%)	4 (13%)	3 (19%)
20-50	61	50 (12%)	7 (23%)	4 (25%)
50+	356	327 (80%)	20 (64%)	9 (56%)
Total	**456**	**409**	**31**	**16**

Table 8: Patients on regualer transfusions, Thalassaemia

Regular Transfusion	Overall	Major	Intermedia	HbEBeta
Yes	393 (86%)	354 (87%)	24 (73%)	15 (94%)
No	65 (14%)	55 (13%)	9 (27%)	1 (6%)
Total	**458**	**409**	**33**	**16**

7.9 Discussion

To summarise, data was lacking regarding transfusion fields. The most valuable data regarding transfusion is from the annual review but as this was recorded in such small numbers and 50% of the data was from one centre it was not analysed. This is because it would have been inherently bias. There have been some assertions made in the analysis but the external validity is poor, particularly in sickle cell disease, as the sample size is small and may not be representative of the population as a whole.

For the future, it may be preferable to record the thalassaemia patients as transfusion dependent or not transfusion dependent. The developing national Haemoglobinopathy genetics database could potentially link to it to provide the thalassaemia genotype. Much data on these patients can be obtained through blood bank manager systems and one could argue that this data should really be electronically transferred rather than asking for manual inputting of data. Much of the lack of data input has been ascribed to the lack of data support. It should be noted that most clinicians and nurses do not have direct access to these laboratory based systems. Lastly there is enormous scope for there to be direct link between the NHS Blood and Transplant data to be linked to NHR. However, much work needs to be done to understand how best that could work to support patients and for the logistics of this. Currently, unless there are particular transfusion requirements, hospital blood banks do not routinely tell NHS Blood and Transplant who the blood they are ordering is for. Ideally, where all this was in place, you would be able to link genotype and other fields in the NHR with transfusion requirement and practice. This would give patients and parents of future patients a much better idea of what the future holds as well as proactively being able to predict and plan for demand to allow for a seamless provision of appropriate blood for patients. All this in a growing patient cohort whose blood needs are not well reflected by the donor demographics where <2% come from ethnic minorities.

CHAPTER 8

Information Service

Mark Foster

8.1 Information Service Overview

The National Haemoglobinopathy Registry Information Service https://mww.mdsas.nhs.uk/nhrinfo allows real time online access to summary information. The flexible reports allow filtering of the Information, allowing users to view the summary data at national level, a regional level and at trust level. The data available through this service is anonymised, real time, single access with the ability to export data locally for analysis.

All users of the NHR are entitled to access the Information Service, and are encouraged to make use of this facility. New reports can be easily added onto the Information Service and MDSAS actively encourage users of the NHR to submit their ideas for reports they believe would add value to the Information Service.

8.2 Reports Overview

The Information Service was developed alongside the NHR, the reports initially made available detailed patient numbers against various indicators such as diagnosis, centre, age group and gender. Figures 8.3 - 8.5 are examples of reports currently hosted on the Information Service. With user requests and new technologies such as mapping becoming available the number of reports available has increased considerably. Examples of new reports available include number of Patients with TCD monitoring and patients treated at London centres living outside of London.

The Information service is to be reviewed in 2015. Reports reflecting the changes in commissioning structure are to be added and any reports deemed no longer of use are to be removed. The layout of the report home screen is also to be reviewed, with different report types grouped together. A brief description of each report will also be added under the report name.

Figure 8.1 Information Service Homepage

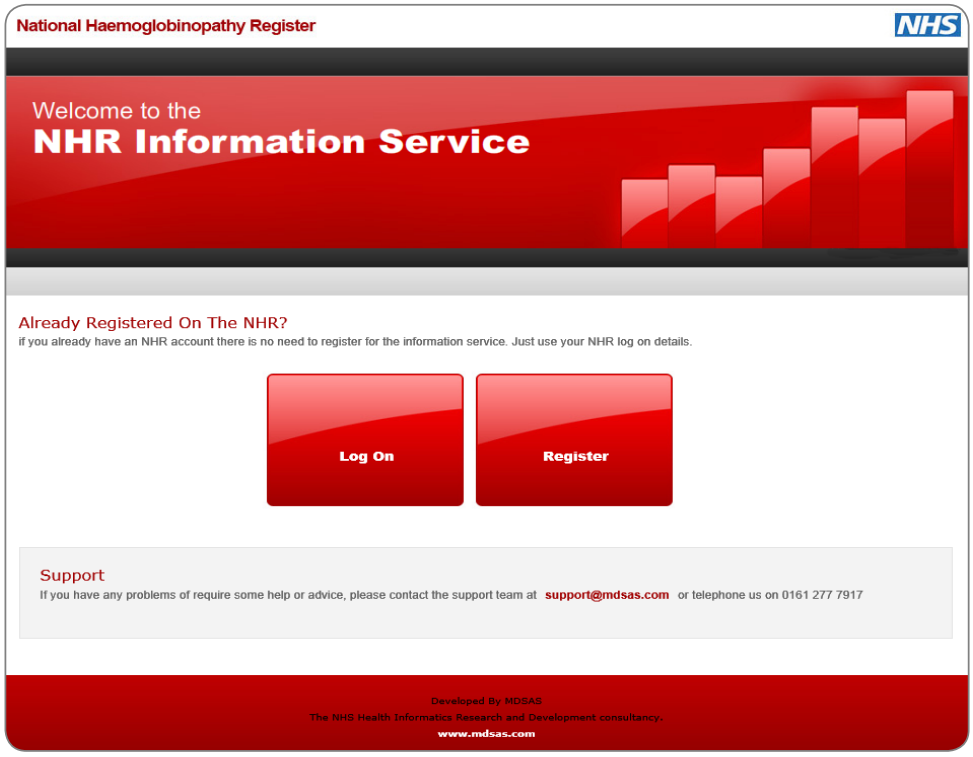


Figure 8.2 Information Service Reports Menu

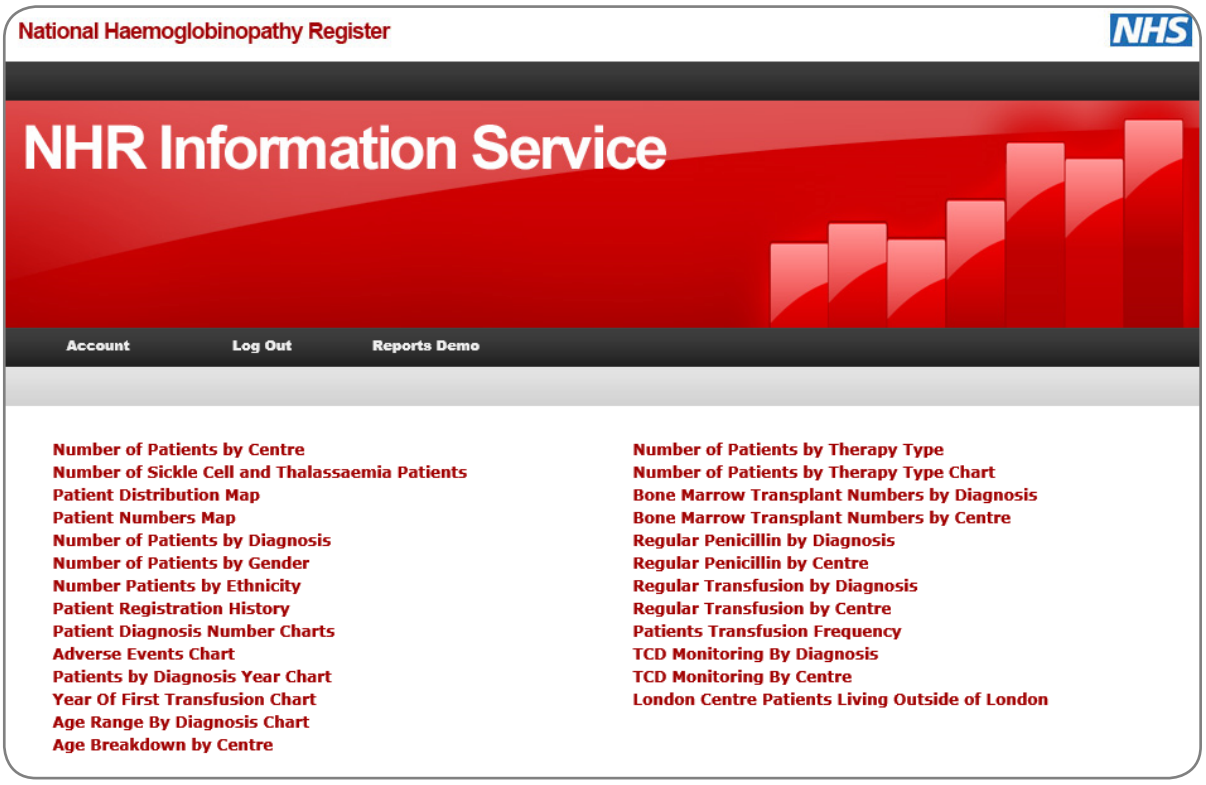


Figure 8.3 Patient Registration History

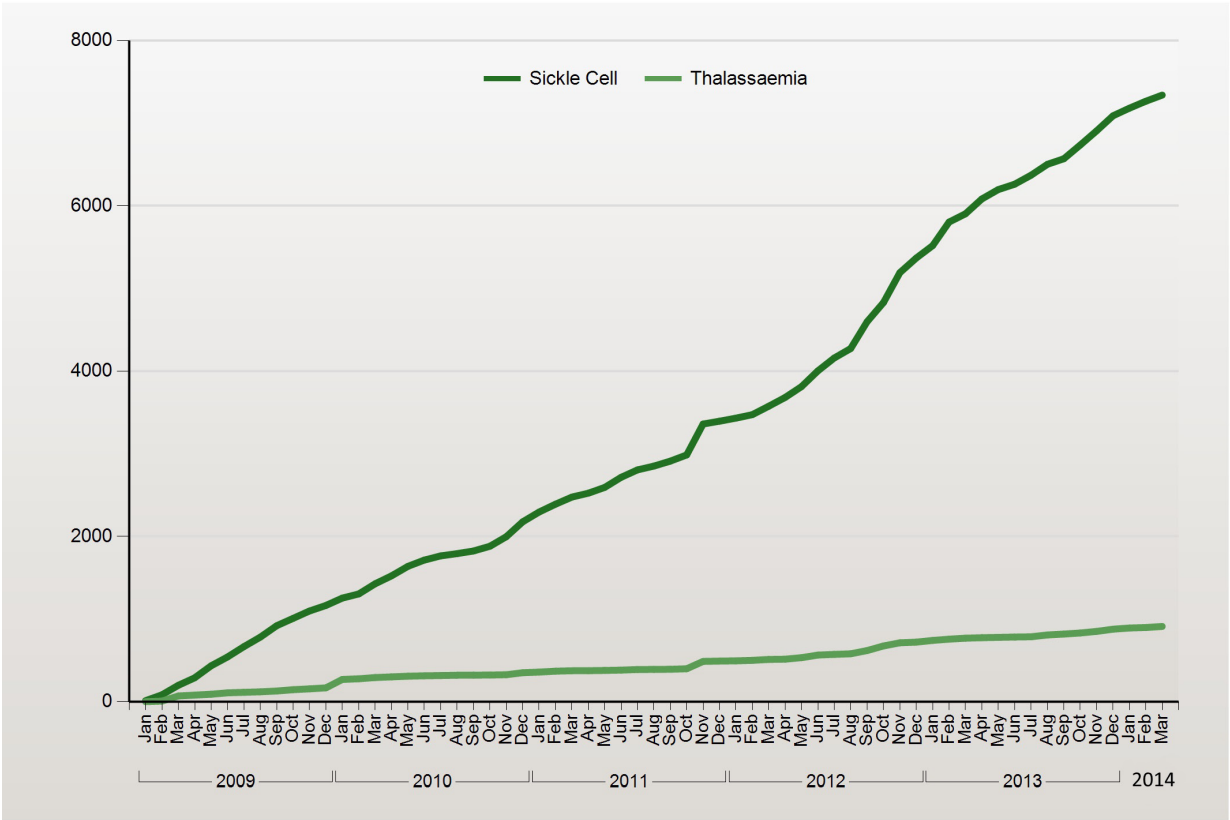


Figure 8.4 Registered patients by Diagnosis & Gender

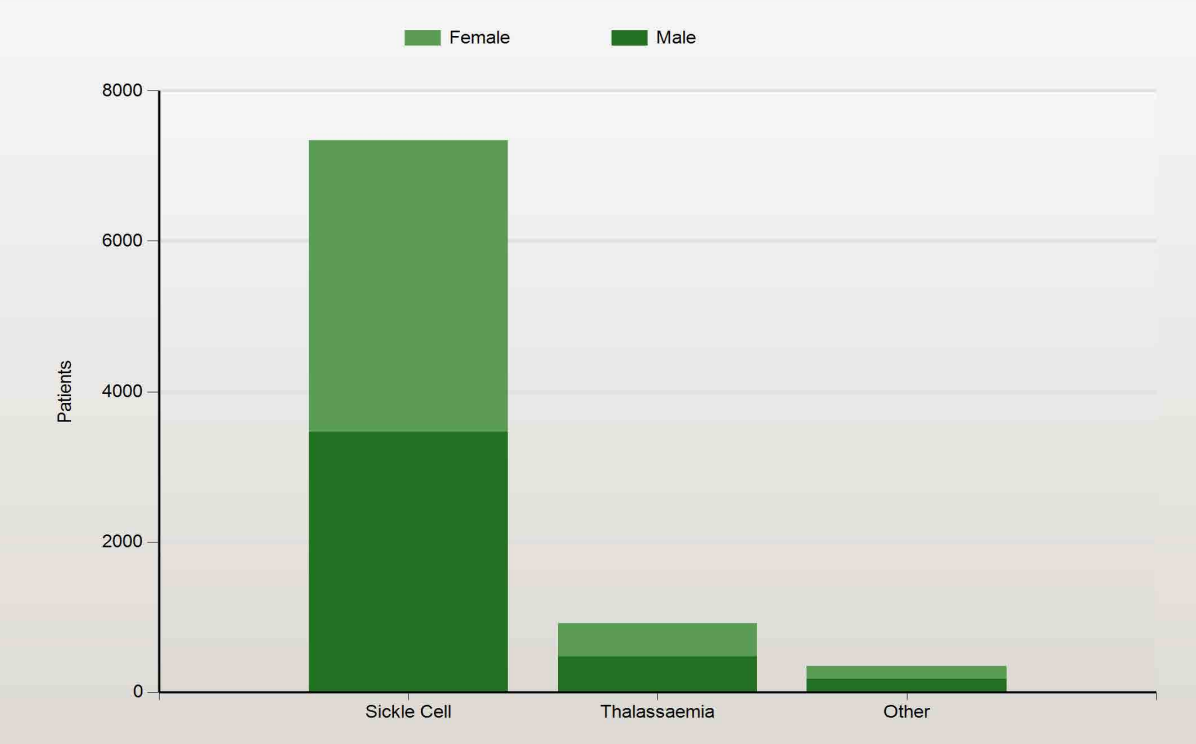


Figure 8.5 Age at Death - All diagnoses

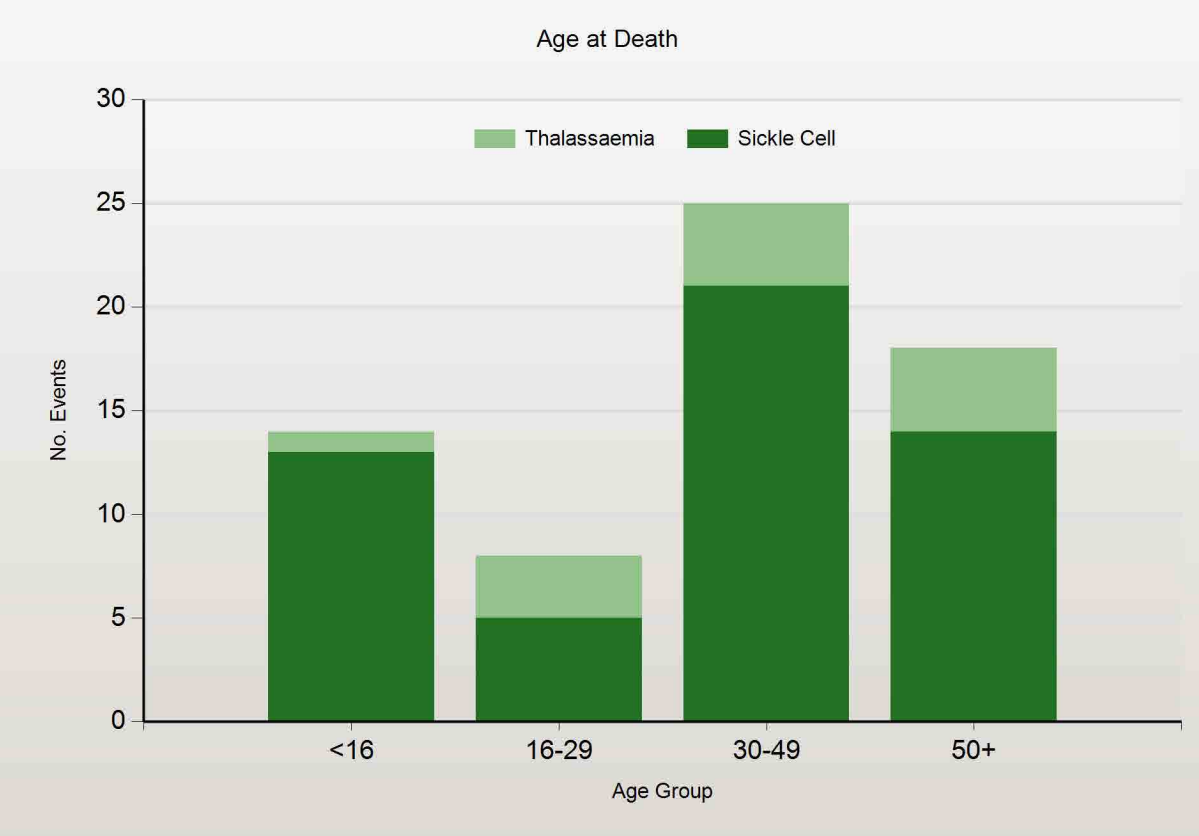
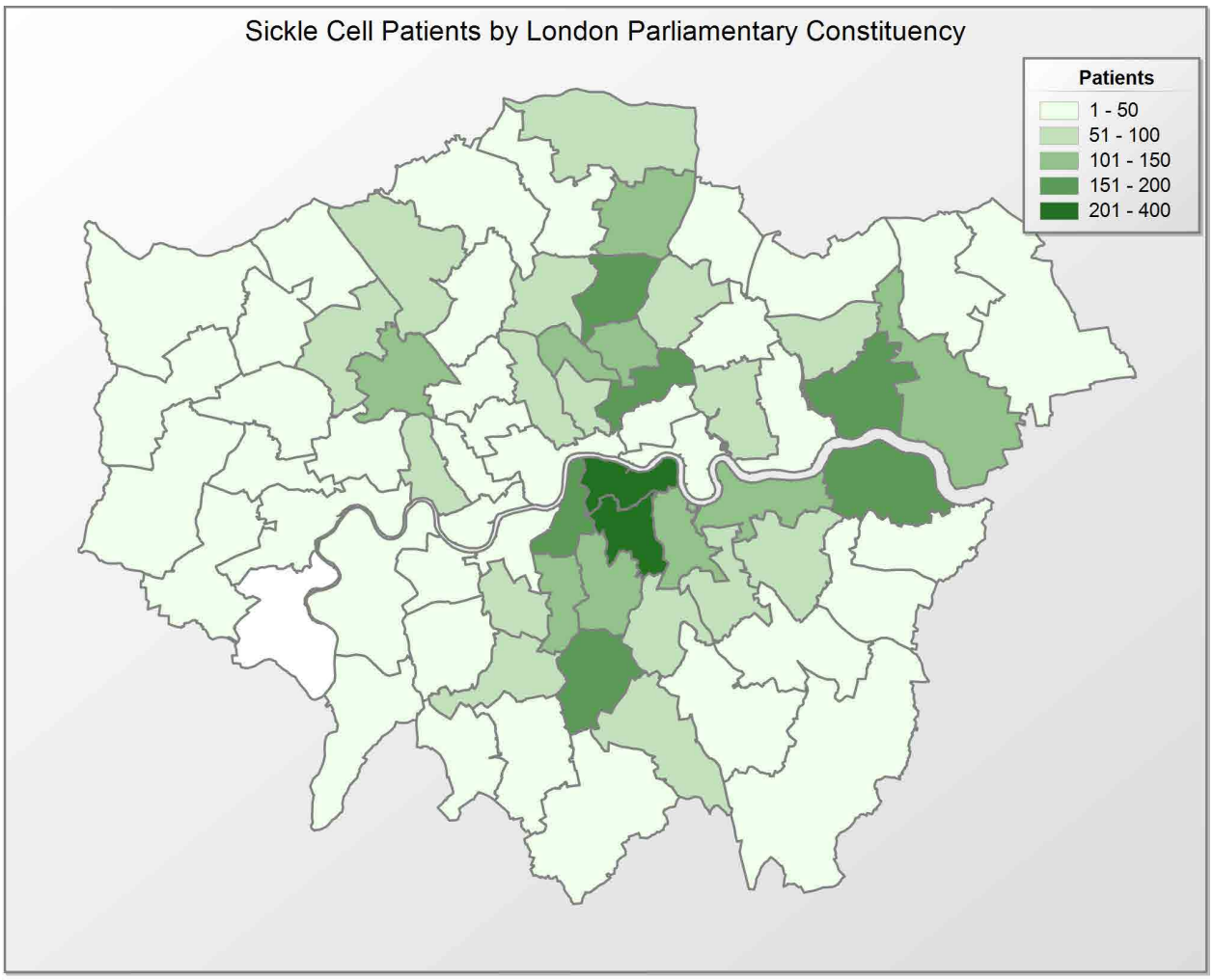


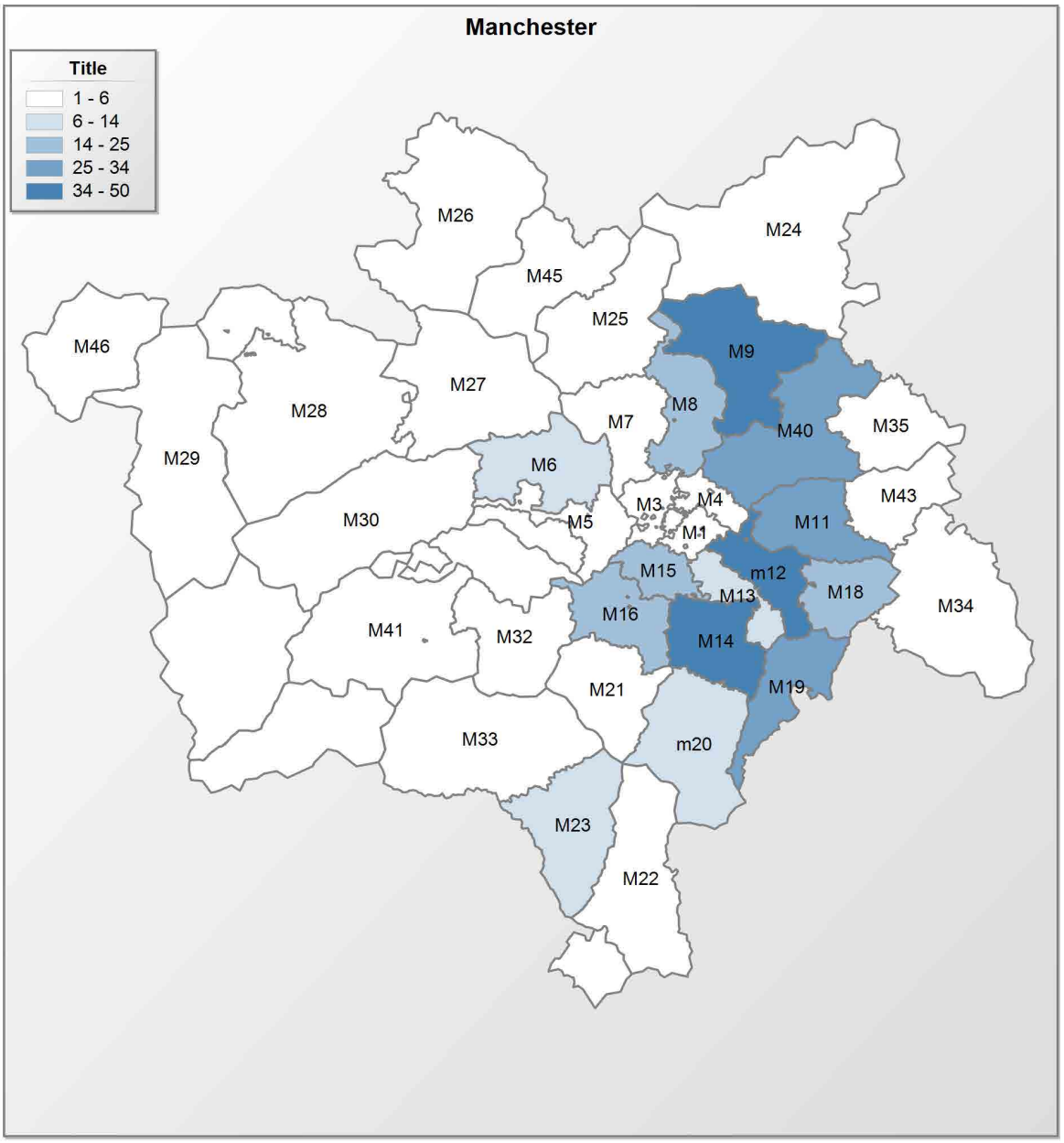
Figure 8.6 Patient numbers by Parliamentary Constituency



8.3 Report Requests

The NHR is with increasing regularity proving to be a valuable source of information for various stakeholders who require information on Haemoglobinopathy patients. The NHR has received information requests in the last year from clinicians, commissioners and political think tanks. Figure 8.6 is a map requested by an organisation called Political Intelligence that details Sickle Cell patient numbers by Parliamentary Constituency. This map was produced with the aim of engaging members of parliament in discussion over the improvement of services for patients with Sickle Cell. Figure 8.7 is a map showing paediatric Sickle Cell patients by Post Code District, this report was requested by a user of the NHR who works with schools educating children with Sickle Cell.

Figure 8.7 Patient numbers by Postcode Disctrict



CHAPTER 9

Adverse Events

Anne Yardumian

9.1 Serious Incident Reporting

In addition to NHR adverse events reporting, all local trust reporting procedures should continue to be followed for the reporting of adverse events.

The NHS 'Serious Incident Reporting and Learning Framework' http://www.nrls.npsa.nhs.uk/report-a-patient-safety-incident/serious-incident-reporting-and-learning-framework-sirl is based on recognition that the benefits from investigating events, usually with a 'root cause analysis' approach, can lead to important improvements in care, and further incidents avoided, resulting in better and safer patient care. We believe that systematic recording of significant complications in our patients, through the NHR - if managed, analysed and communicated appropriately - should have considerable value in future so that:

- If, on review of a serious event in a patient at any particular site, some aspects of care which might have contributed to a complication or poor outcome are identified and it was possible to learn ways in which management might be bettered for a similar case in the future, this learning can be shared by the professional community to enable other clinicians to better manage similar problems in their own patients
- To enable the occurrence of an unusual serious event or complication, which might be seen in a different centre in the near future, to be circulated to clinicians to forewarn them of a possible local occurrence. In this way doctors and nurses can be made alert to a similar problem in any patients under their care, and can diagnose and treat promptly, giving the patient the best chance of recovery.

It is clearly important for patients and families, as well as healthcare professionals, to know

- The current incidence of key complications
- Life expectancy and causes of death for these patients in England today, about which we are currently unaware.

Finally, in addition to the negative impact on patients, the consequences of serious events, such as emergency admissions or ITU stays, can place a significant financial burden on the NHS potentially taking money away from other areas of patient care. Any reduction in the number of serious events as a result of this initiative will help drive up quality, improve the patients' experience and reduce costs to the NHS.

PLEASE NOTE: the responsibility for reporting a Serious Incident externally in the usual way, in addition to NHR reporting, remains and is the responsibility of the Trust in which the incident occurred.

To be maximally useful, there will need to be a way of communicating with all red cell clinicians, not just those entering patients onto the NHR – although with time it is hoped this would include most of those treating patients.

It has been suggested that the term 'adverse event' may not be appropriate; the term is widely used, but usually with the implication that the issue was a problem [avoidable or not] in care rather than a problem associated with the condition itself. Certainly some of those we see ARE attributable to the condition and not to any aspect of care. We might instead refer to **'serious events'**

9.2 Process Review

We have, at the UK Forum on Haemoglobin Disorders Committee and at the Clinical Reference Group meetings, taken the opportunity to review the current process, and to recommend the following changes and improvements to process.

1. There should be a mandatory box on the reporting page to give the underlying haemoglobin disorder diagnosis; this should be by subtype of sickle cell disease or thalassaemia.
2. The date field should allow reporting only back to, say, 6 months previously: this should allow time for case review / p.m. findings but allow a manageable number of usefully recent cases to be reported for analysis and communication.
3. The categories of events to be reported ;

a. Death

b. Bacteraemic sepsis , please specify nature of infection

c. ICU admission

d. Stroke

e. Cardiac dysfunction

f. ESRF requiring dialysis

g. Postoperative complications: ACS, postoperative painful crisis, thrombosis, postoperative bacterial infection, death, readmission within 30 days.

h. Complications during and after pregnancy , IUD , premature delivery < 36 weeks gestation , post-delivery ACS

i. Hyperhaemolysis

j. Other - i.e. any event Sickle / Thalassaemia or treatment related which causes significant long term morbidity.

It is recognised that 'other' may cause some centres to report clinical events which others may not; however, in the interests of getting full reporting on those events we would definitely want to know about, it is probably better to allow reporting of anything reporters feel warrants inclusion. There should be a comment noting that there will be further communication about serious events, and those that are less clinically severe or significant will be included, by group, in each summary or overview report [see 10].

4 The 'free text' box should prompt to give a brief account of event, to a maximum of 500 words.

5 There should be a field on the reporting page, after the free text box asking 'was this event discussed at a mortality / morbidity review meeting: yes / no'.

If yes: opens another box asking : 'please indicate any learning points which arose from this case, e.g. failure of prophylaxis or screening, delays in presentation, identification or intervention, or any other points of discussion which might inform future practice'

If no: this should automatically ping an e-mail back to the person reporting, requesting a team review of case, looking for the same points and requesting entry of any findings by a certain date.

6. In any case, there will be an auto response saying 'thank you for reporting this serious event which has occurred in one of your patients. A small team of specialist healthcare professionals will ensure that your case and your review findings are included, anonymously, in their summary report which you will automatically receive'. This assumes that summary report will be sent out to all those who register patients on NHR – at least, and to other relevant clinicians when we agree a mechanism to reach them.

7 The case reports, after entry onto the NHR, should be sent to a group of three healthcare professionals: ideally a paediatrician or paediatric haematologist, an adult haematologist, and a specialist nurse. One or more should be members of the CRG. Others – for example a biomedical scientist, or TCD expert, may be co-opted for consideration of particular events. They might be asked to undertake this work for a period of 2 – 3 years, with 'staggered' replacement so that some experience can be handed on rather than replacing the whole team at the same time.

8 The NHR should communicate reports to this core team with the reporting centre of hospital encoded. The core team members will need to know only the code for their own centre, so that they can exclude themselves from consideration of any events reported by their own geographical area. For considering events reported from the Centre / linked hospitals of one of the core team, a member of the CRG should be asked to substitute.

9 It is suggested that this core team be appointed after consideration of applicants

an invitation by the CRG Chair to the UK Forum on Haemoglobin Disorders membership. They will be accountable to the CRG for this work, and any reports etc. should be seen first by the CRG before wider dissemination. While the names of those on the clinical panel need not be included, the fact that the event report is to be sent only to 3 clinicians working in the clinical area, and their respective disciplines, should be visible on the NHR reporting page for transparency. Responsibility for data quality and confidentiality would rest with the NHR management team.

10 The three clinicians on the Panel should check event reports as they come in, and

a] communicate among themselves and then with CRG Chair if they feel there are specific concerns, for example a run of similar adverse events from the same Centre, which might prompt immediate further discussion / investigation with the clinical team. The CRG Chair will decide what, if any, early action is needed. While the purpose of reporting is to learn and NOT to censor or judge performance, there needs to be a mechanism whereby an event / a number of reported events, which give reason to question care quality at a given provider site, can be followed up.

b] Otherwise collate them and produce a summary or overview annual report. This will be anonymised [removing any identifiable patient or centre details] and include the numbers of different types of event, by ages and diagnosis, and any lessons learned on case review.

c] Write the reports in such a way that it will be suitable / comprehensible for patients and user groups as well as healthcare professionals.

11 Summary reports will be sent automatically to all those who register patients on NHR, to members of the UK Forum on Haemoglobin Disorders, and members of the CRG. They will also be made available to voluntary organisations and user groups, and Commissioners.

Figure 9.1 Adverse Events Flowchart

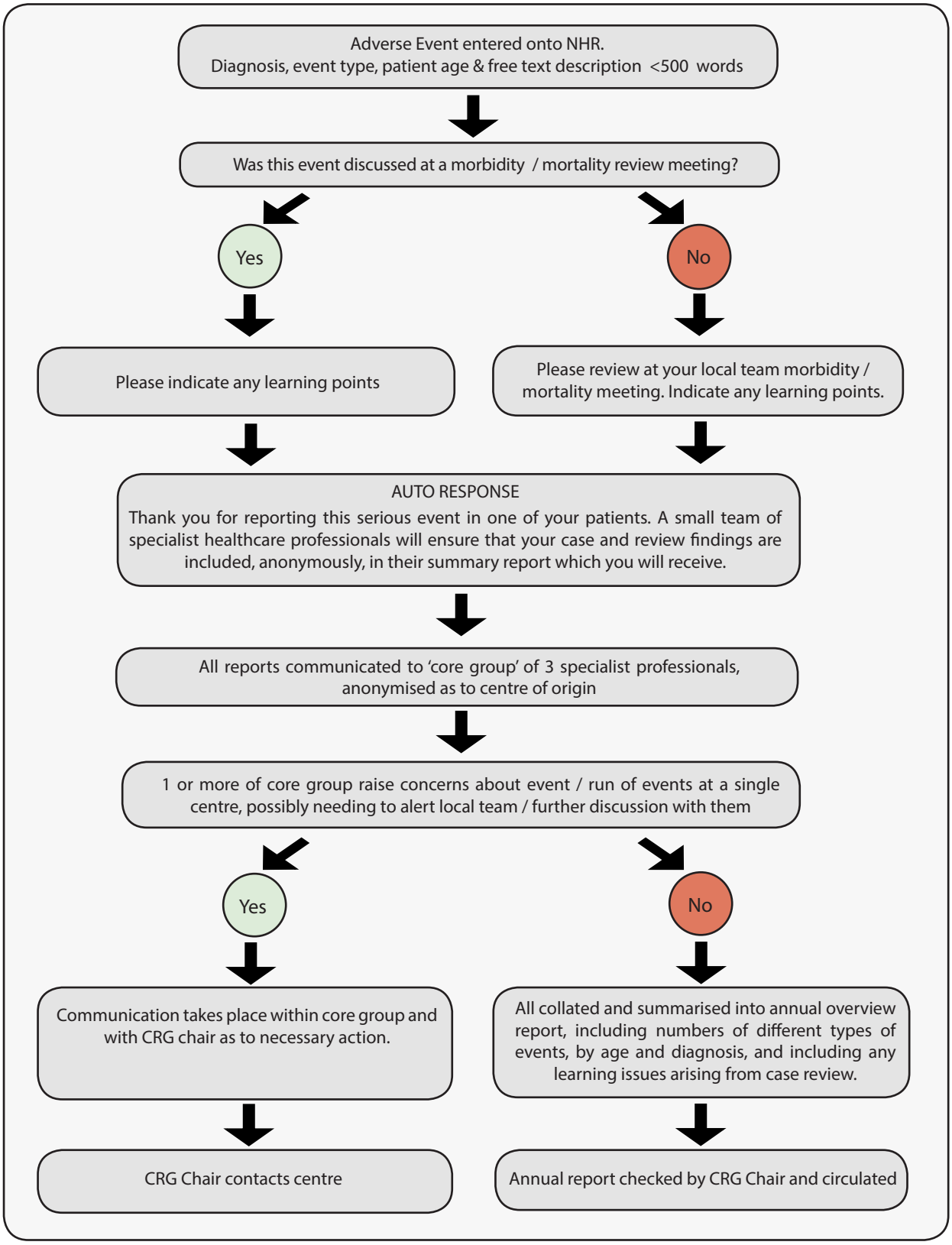


Figure 9.2 Reported Adverse Events History

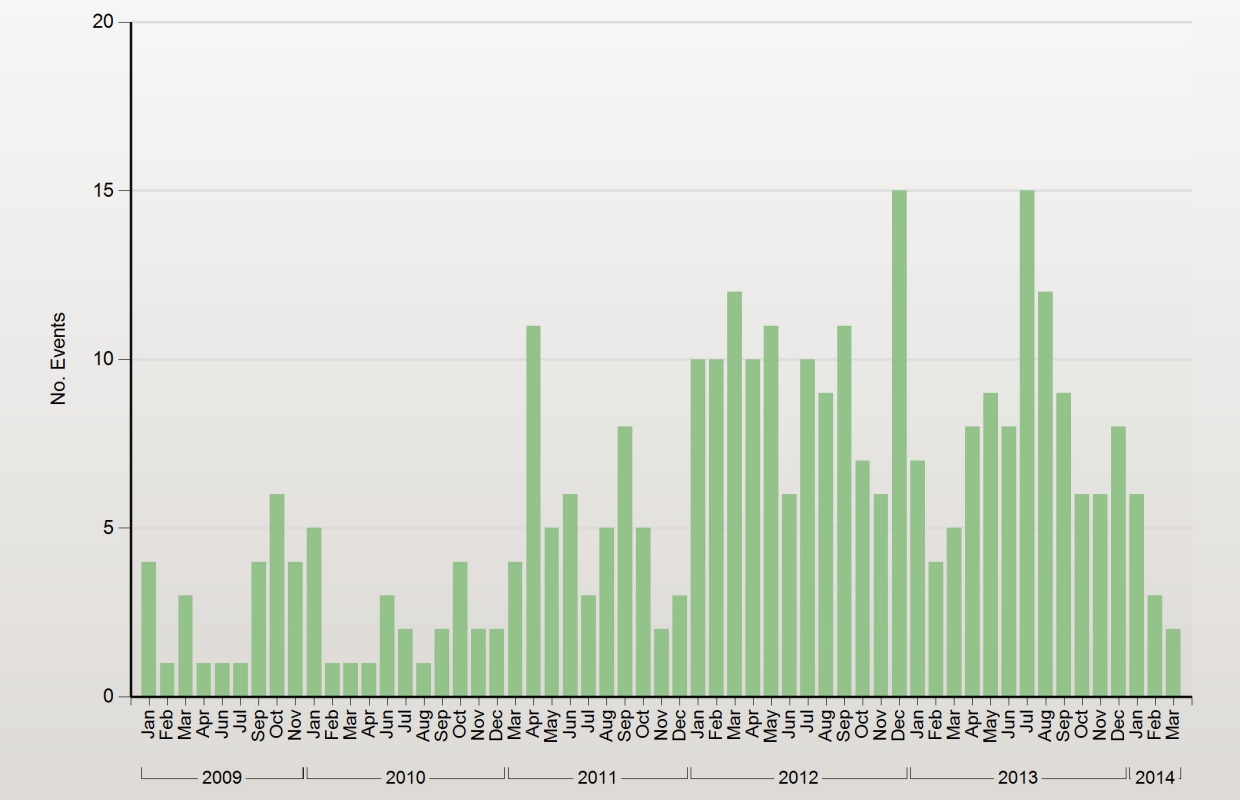


Figure 9.3 Adverse Events - Sickle Cell

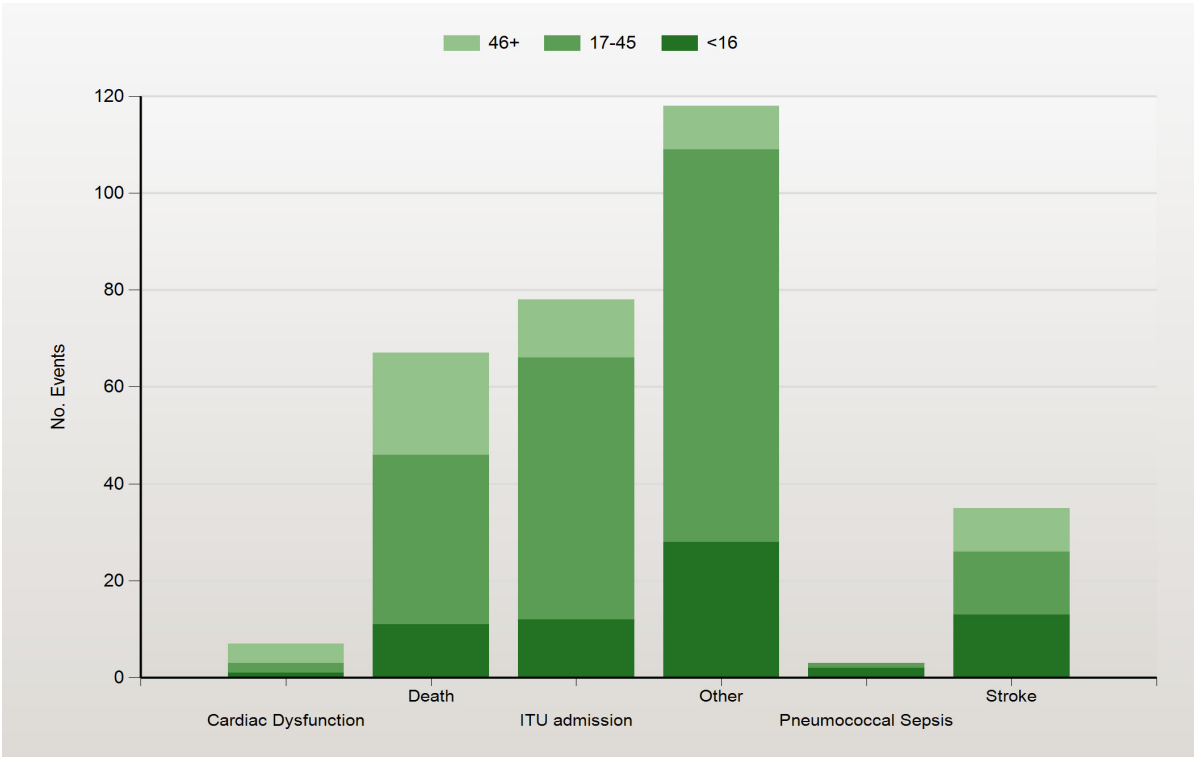
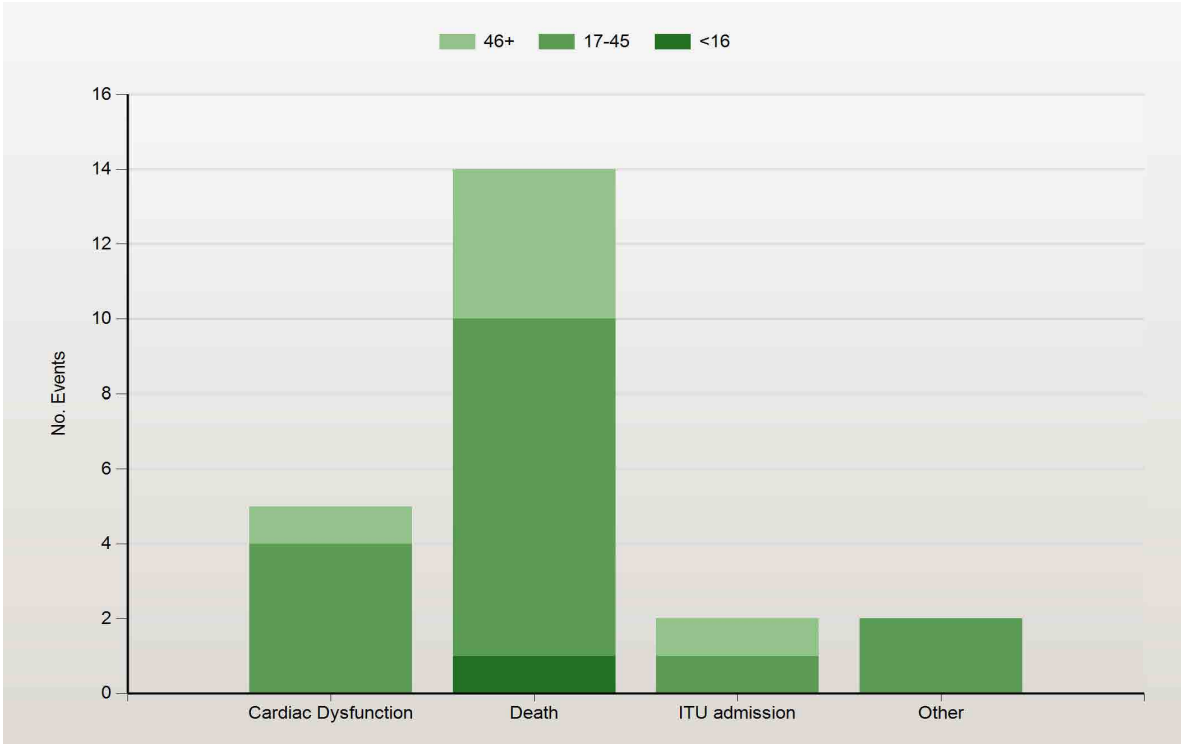


Figure 9.4 Adverse Events - Thalassaemia



CHAPTER 10

Annual Review

Jo Howard

10.1 Role of Annual Review

The Annual Review has been highlighted as a key part of service provision in the specialist service specification. The Annual Review allows a formal assessment of the patient's clinical care and is especially important for patients with complex care needs. An annual review will include:

- A review of acute complications to ensure they are being appropriately managed and that acute pain management protocols are adequate
- A review of chronic complications to ensure they are being monitored and treated appropriately and that specialist referrals have been made if necessary
- A review of screening investigations including Trans Cranial Dopplers in children and echocardiography or ophthalmology screening in adults
- Discussion of self-care, including reproductive options and partner screening if appropriate, and alcohol/smoking
- A review of medications and vaccinations
- A discussion of disease modifying therapy e.g. hydroxycarbamide, transfusion
- A discussion of local research studies or recent advances in care which may alter management
- An opportunity for the patient to ask questions about their care

All patients with Haemoglobinopathies should have a comprehensive annual review performed by their specialist centre, or accredited local centre. These are best performed using a standard pro forma and several units across England have developed these. The NHR pro forma has tried to develop a minimum data set, so that basic information can be collected on every patient but allows local expansion if centres wish to collect more information. The information from the Annual Review can be printed out and given to patients, GP and local care givers and provides a summary of current issues, or it can be used as the basis for a care plan.

The use of the NHR as a tool for Annual Review has several advantages:

• Provision of up to date patient numbers from each centre. The Haemoglobinopathy peer review found that very few centres had accurate up to date patient numbers. This is a very mobile population with a large number of transient patients. Therefore patient lists often included 'all patients seen in the last 2-5 years' or, 'all patients with any contact with service'. This leads to over counting and double counting of patients, who are often attending several centres and included in the figures at all centres. The use of the annual review means that the centre has accurate figures on the numbers of patients who are engaged with outpatient services and who are receiving comprehensive care there.

• The ability to track patient numbers over successive years. There is little evidence of capacity planning for Haemoglobinopathy services across England and there are several centres with rapidly enlarging population. Accurate data collection is necessary to plan for these increasing patient numbers and implement growth of services in the areas where they are most needed and the annual review functions will provide this data.

• The NHR allows data to be collated from the Annual Review returns. This makes it very straightforward for centres to obtain data on

1. Numbers of patients with each diagnosis (SCD or Thalassaemia).
2. Age break down of patients, which is very important for service planning, in particular for transition services.

3. Numbers of paediatric patients who have had a trans-cranial Doppler (TCD) performed, and the results of this study. This is a key Quality Standard Indicator and the NHR annual review has the ability to collect this information as a report. It can also be used to produce a list of patients who have had a TCD to highlight the patients who still require this investigation.

4. Numbers of patients receiving chronic blood transfusion therapy and iron chelation therapy. Accurate figures on patients receiving blood transfusion therapy and types of chelation therapy is not easily available at the majority of centres, but this is a key area for both quality improvement and service planning. The NHR Annual Review allows accurate data to be obtained on numbers of patients being transfused, method of transfusion and type of iron chelation and efficacy of iron chelation (by reviewing markers of iron overload)

The data can be used by each centre to look at its own patients, review adherence to quality standards and to obtain useful data on growth of the service. The data can also be used in a comparative way across England to assess overall quality of care, or can be collated nationally to show trends in care, or highlight concerns across the whole country.

The Annual Review is used very successfully in other specialties, including haemophilia. The information collected from Annual Review in these services is used successfully to review quality of care, compare care in different centres and to help centres to improve quality of care. It also has a vital role in service planning and to ensure that resources are being used most appropriately. Haemophilia Centres already use the NHR to perform their Annual Reviews and this is used to produce an Annual Report. This provides a good example of the use of the NHR Annual Review and examples of useful data fields that apply to Haemoglobinopathies as well as haemophilia include:

• Cause of death reporting

• Complication rate reporting – bleed rate and rate of inhibitors are collected in the haemophilia population, but in the Haemoglobinopathy population, in addition to information reported separately via the adverse event reporting, information on pregnancy rates or incidence of surgery and surgical complications could be collected

• Use of treatment – in haemophilia this applies to factor usage, but in Haemoglobinopathies could be applied to use of iron chelation or hydroxycarbamide

• The numbers of patients treated with factors per centre per year is collected for haemophilia, similarly the numbers of patients receiving blood transfusion or who have necessary investigations such as trans-cranial Doppler USS could be collected.

10.2 Data from Annual Review

The Annual Review function has been in place since 2011/12 But has only been used by a minority of centres, in part because of a lack of administration support and in part because functionality had not been optimal. In 2012/13 1017 patients had an annual review performed. 19/48 registered centres were performing Annual Reviews using the NHR and in these centres the percentage of patients receiving an Annual Review on the NHR ranged from 2% to 92%.

10.3 Updates to Annual Review

During 2012/13 the Annual Review has been updated and the format has been changed to improve the ease of usage. The Annual Review screens were updated following extensive input from the NHR steering group. Following this update there has been further input from users and the steering group about the functionality of the Annual Review and it is currently undergoing further revisions. These will be implemented by MDSAS and updates will continue on an annual basis.

10.4 Summary of changes to Annual Review

Patient details: This provides an opportunity to change any patient details and record basic information on hospital admissions, transfusions, surgery and pregnancy in the last 12 months
'History' button is to be added to show previous entries of the Annual Review. There is also going to be opportunity to record that a patient was offered an annual review but did not attend
Therapy section: This allows recording of patients on transfusion and iron chelation therapy. Hydroxycarbamide therapy is currently recorded here but is going to be moved to the medications section
Medications and Vaccinations section: To record Medications and Vaccinations
Investigations: To record routine blood tests and other screening investigations. Observations (Blood pressure and oxygen saturations) are currently in this section but are to be moved to the patient details section.
Trans-cranial Doppler has its own section. Specialist imaging (Cardiac T2* and Liver iron concentration – Ferriscan) is to be moved to its own section
Operations: Records any operations performed in the previous 12 months
There is a free text section to allow additional information to be added

10.5 Future of Annual Review – fitting in with Quality Standards

The Clinical Reference Group (CRG) is working with NHS England to develop a Quality Dashboard. This will include data which will need to be reported to specialist commissioners on a quarterly basis, to allow them to review adherence to quality standards and ensure quality of care. The majority of this data can be produced automatically from the Annual Reviews performed on the NHR.

The measures that will be collected on the Quality Dashboard have yet to be confirmed by the Clinical Reference Group and NHS England, but they are likely to include:

- Outcome of defined surgical procedures within network
- Number of children within at risk group receiving annual Trans cranial Doppler screening
- Number of children identified by neonatal screening who enter care pathway
- Number of children beginning Penicillin at or before 3 months
- Number of Annual Reviews performed
- Number of sickle cell patients receiving long term top up or exchange transfusion
- Number of patients on long term transfusion receiving iron chelation and the adequacy of the iron chelation therapy
- Number of patients on Hydroxycarbamide therapy

There are a few elements of the Quality Dashboard which will not be able to be collected from the Annual Review these include:

- Adverse events entered on the NHR
- Timeliness of pain relief in SCD

The use of the NHR Annual Review will therefore aid clinicians in reporting the majority of elements on the Quality dashboard. In the future if these quality requirements change it will be possible to build them into the Annual Review NHR function.

CHAPTER 11

Developing the NHR

Kate Ryan

11.1 Future developments

As the NHR continues to grow in strength, a number of developments are planned which will enhance the benefits for both clinical staff and patients. Some of these developments are very close to fruition while others are at earlier stages of discussion.

11.2 Patient cards

A centre will be able to print out a wallet-sized card, which can be carried by the patient containing important personal information. The card can be printed using standard printers in a patient's centre onto an information sheet. The card will contain important details of diagnosis, contact details for that patients registered centre and highlight significant clinical issues around transfusion or complications. An example card is given below:

NATIONAL HAEMOGLOBINOPATHY PATIENT CARD

Name:	Test Patient		
Date Of Birth:	01/01/1990	**NHS No:**	
Diagnosis:	Sickle Cell Anaemia (Hb SS)		
Red Cell Antibodies:	No		
Local Hospital Team:			
Specialist Centre:	TEST CENTRE		

*Contact Specialist Centre (0161 123 4567) for clinical or transfusion advice.

11.3 TCD Quality assurance

Transcranial Doppler (TCD) screening predicts children with sickle cell disease who are at risk of stroke and should be offered to all children as standard care. An abnormal TCD triggers a pathway of further investigation and management which may results in long term transfusions. Although TCD is offered in many centres there has not been a recognised quality assurance scheme to ensure consistent and accurate results throughout the UK. It is proposed that the NHR acts as a repository for scans which have been undertaken in local centres and that these scans can then be directed for expert independent analysis. Details of this scheme are still under discussion.

11.4 User involvement

The NHR steering board is keen to promote greater user interaction and is seen as a way to engage with patients in their home environment away from the hospital clinic. There are a number of examples of how this might benefit:

I. Patient surveys: Feedback from patients is a key component to shape and improve services. Experience from the peer review programmes highlighted the difficulties faced in conventional paper based postal surveys of face to face meetings where limited responses do not necessarily reflect all the patients' experiences. The user group of the CRG is developing patient feedback questionnaires and it is hoped that these could be completed electronically and collated by the NHR

II. Links to information and education such as UK Forum website, NICE, and location and contact details for specialist centres.

III. Direct input into NHR records by patients. This could be potentially useful for recoding painful crises or other significant patient clinical experiences which could then be reviewed in the outpatient clinic.

11.5 Using the NHR as a tool for epidemiological studies

The NHR offers a unique opportunity to examine the natural history of Haemoglobinopathies in a universal health care system. Information on outcomes and complications based on published case series may no longer be accurate with modern management and therapies. For instance, life expectancy is unknown and there are now considerable numbers of older patients registered on the NHR. Furthermore the impact of other conditions is also unclear. Knowledge of mortality and morbidity may usefully direct specific areas for research. Proposals for the interpretation of epidemiological data on the NHR will be carefully considered by the steering committee with the involvement of users.

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