# National Haemoglobinopathy Registry

## ANNUAL REPORT 2022/2023

# Contents



***Figures***



***Tables***

# Chapter 1: Introduction

The 2022/23 Annual Data Report provides stakeholders of the NHR with an update on patient numbers for Sickle Cell , Thalassaemia and other rare inherited anaemia patients in England.

The removal of inactive patients from reporting is necessary to maintain an accurate portrayal of patient populations.

| Status | No. |
|---|---|
| Deceased | 71 |
| Emigrated | 12 |

Table 1 - Inactive Patients details the number of patients that have become inactive during the year. We encourage users of the NHR to update the status of their patients accordingly.


| Status | No. |
|---|---|
| Deceased | 71 |
| Emigrated | 12 |


*Table 1 - Inactive Patients.*

# Chapter 2: Sickle Cell Reports

| SHT Name | Registrations |
|---|---|
| Barts Health NHS Trust | 1674 |
| Birmingham Women's and Children's Hospital NHS FT and Sandwell and West Birmingham Hospitals NHS Trust | 1341 |
| King's College Hospital NHS Foundation Trust | 1333 |
| Guy's and St Thomas' NHS Foundation Trust | 1285 |
| North Middlesex University Hospital NHS Trust | 705 |
| Lewisham and Greenwich NHS Trust | 692 |
| University College London Hospitals NHS Foundation Trust | 640 |
| Manchester University NHS Foundation Trust | 639 |
| St Georges Healthcare NHS Foundation Trust | 594 |
| Imperial College Healthcare NHS Trust | 527 |
| London Northwest University Healthcare NHS Trust | 508 |
| Oxford University Hospitals NHS Foundation Trust | 468 |
| Leeds Teaching Hospitals NHS Trust | 416 |
| Whittington Health NHS Trust | 395 |
| University Hospitals of Leicester NHS Trust | 384 |
| Homerton Healthcare NHS Foundation Trust | 366 |
| Croydon Health Services NHS Trust | 296 |
| Nottingham University Hospitals NHS Trust | 275 |
| University Hospitals Bristol & Weston NHS Foundation Trust | 224 |
| The Newcastle Upon Tyne Hospitals NHS Foundation Trust | 194 |
| Addenbrooke's Hospital Cambridge (Cambridge University Hospitals NHS Foundation Trust) | 178 |
| Royal Liverpool University Hospital (Liverpool University Hospitals NHS Foundation Trust) | 171 |
| Sheffield Teaching Hospitals NHS Foundation Trust | 124 |
| University Hospital Southampton NHS Foundation Trust | 116 |
| Sheffield Children's NHS Foundation Trust | 71 |
| University Hospital of Wales (Cardiff and Vale University Health Board) | 56 |
| Sandwell Hospital - Paediatrics | 11 |
| No SHT specified | 177 |

*Table 2 - Sickle Cell registrations by SHT.*

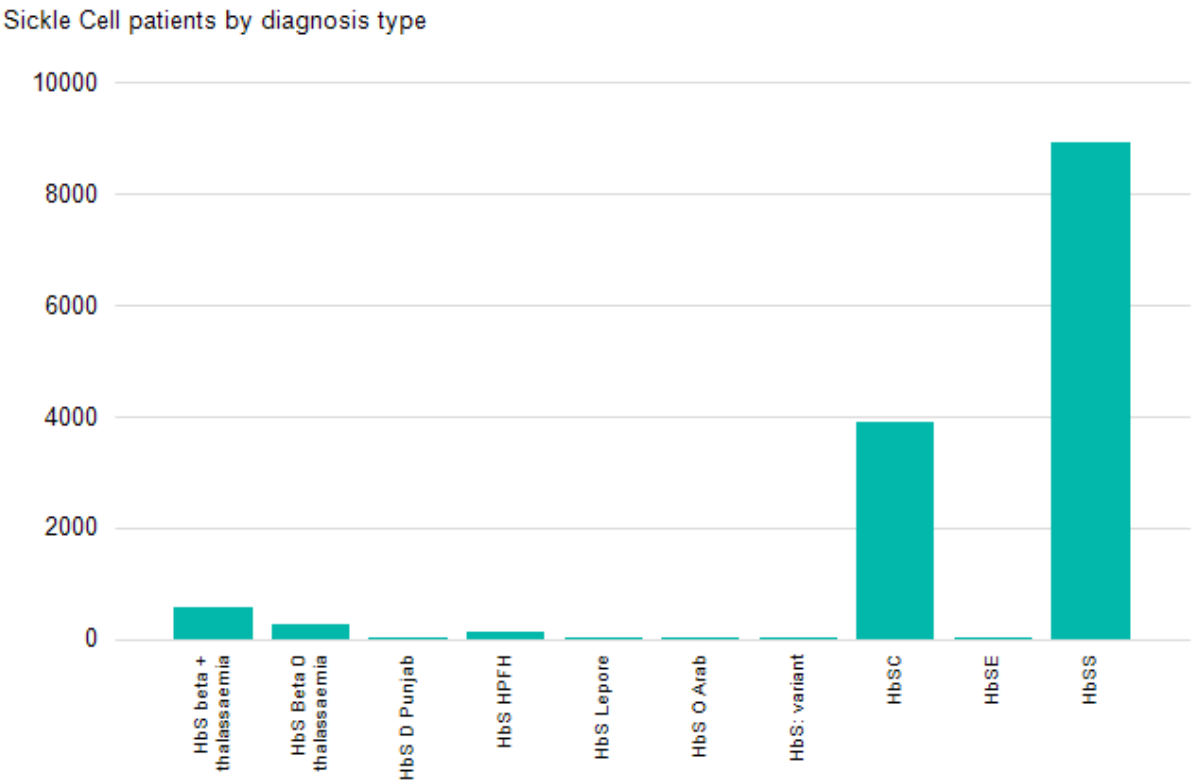


*Figure 1 - Sickle Cell patients by diagnosis type.*

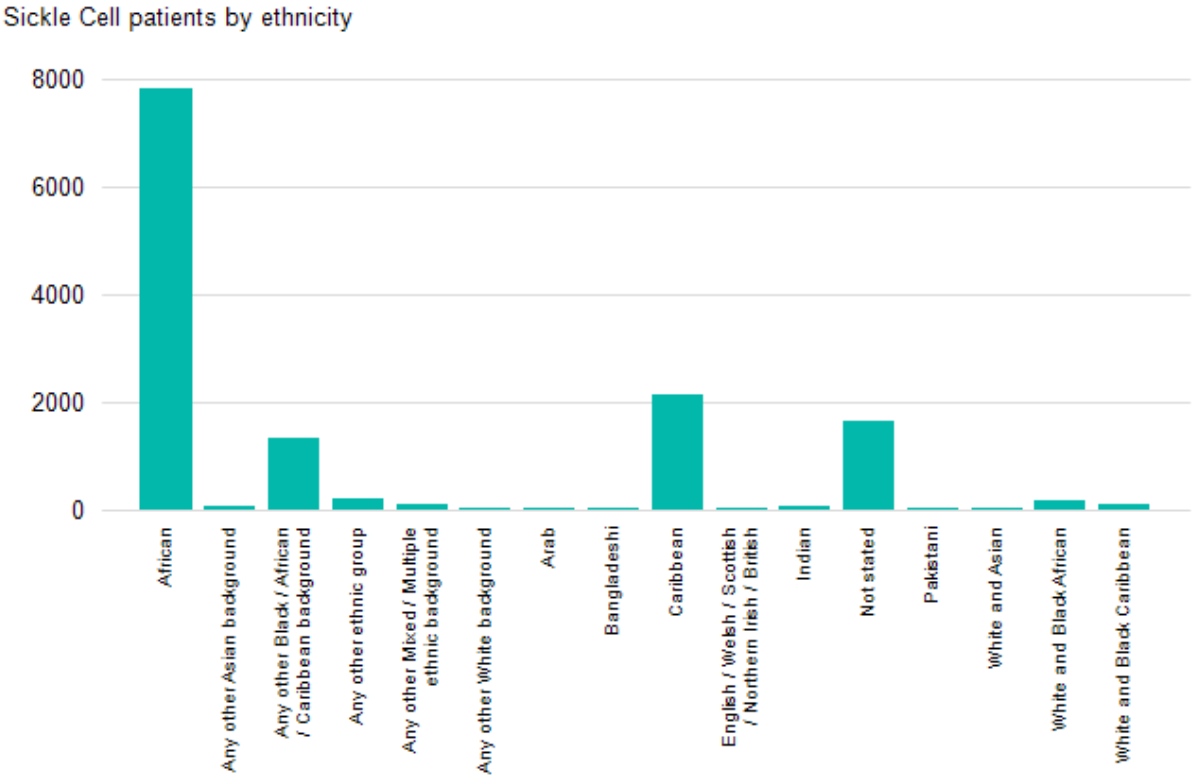


*Figure 2 - Sickle Cell patients by ethnicity.*

| HCC Name | Registrations |
|---|---|
| South East London and South East | 3618 |
| East London and Essex | 2063 |
| North Central London and East Anglia | 1893 |
| West London | 1666 |
| West Midlands | 1349 |
| North East and Yorkshire | 812 |
| North West | 805 |
| East Midlands | 654 |
| Wessex and Thames Valley | 584 |
| South West | 229 |
| London and South East | 5 |
| London, South Central and South West | <5 |
| Midlands | <5 |
| North | <5 |
| No HCC specified | 176 |

*Table 3 - Sickle Cell registrations by HCC.*

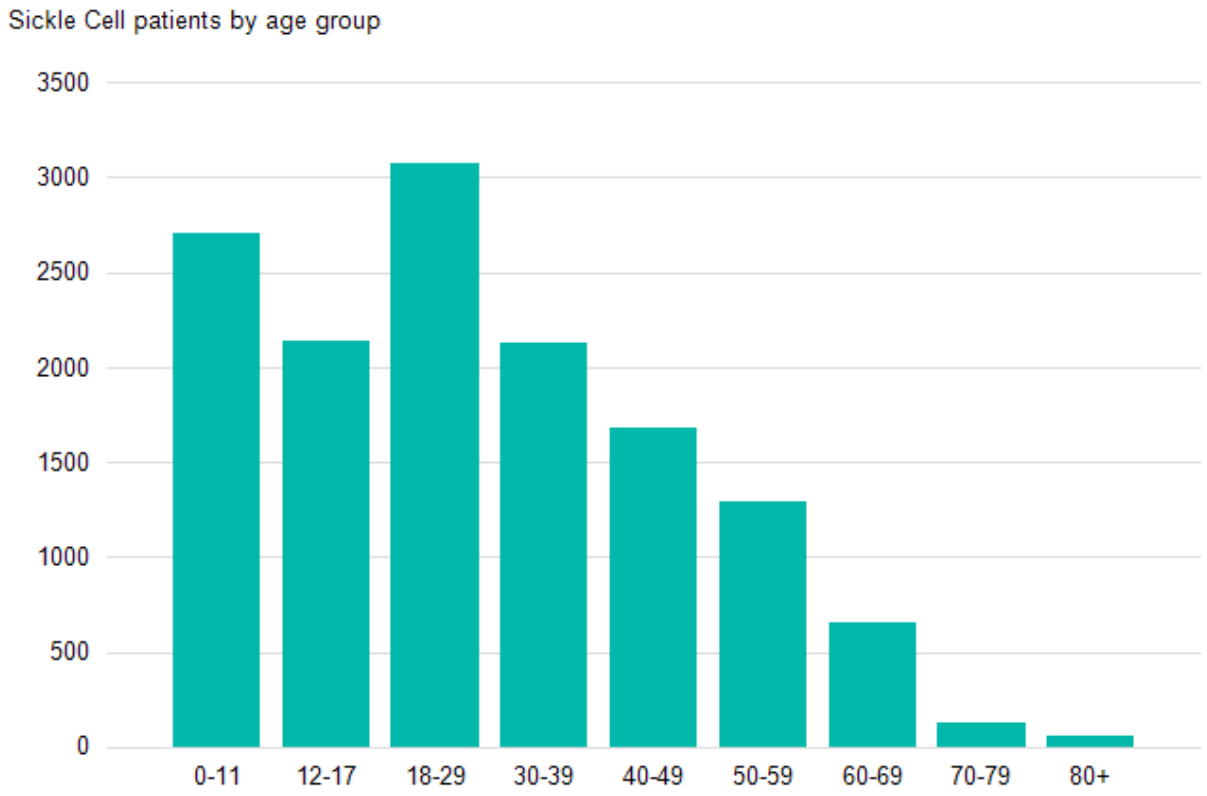


*Figure 3 - Sickle Cell patients by age group.*

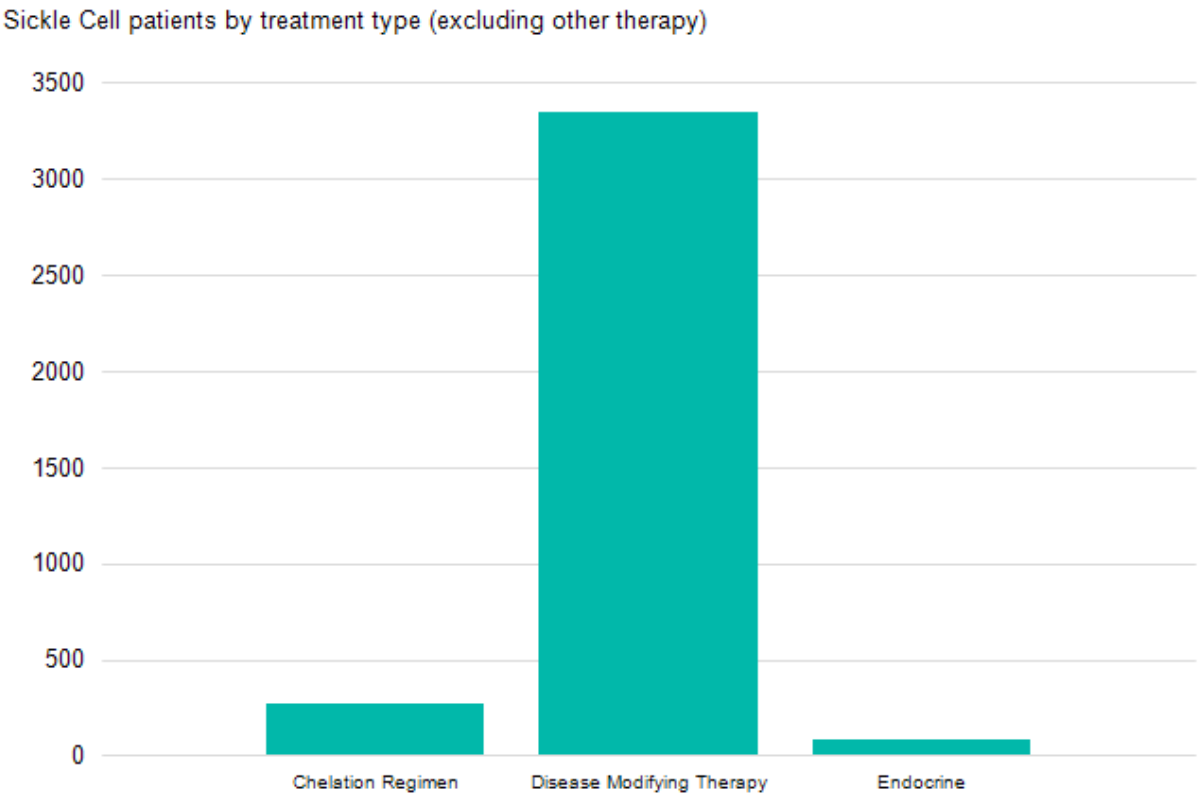


*Figure 4 - Sickle Cell patients by treatment type (excluding other therapy).*

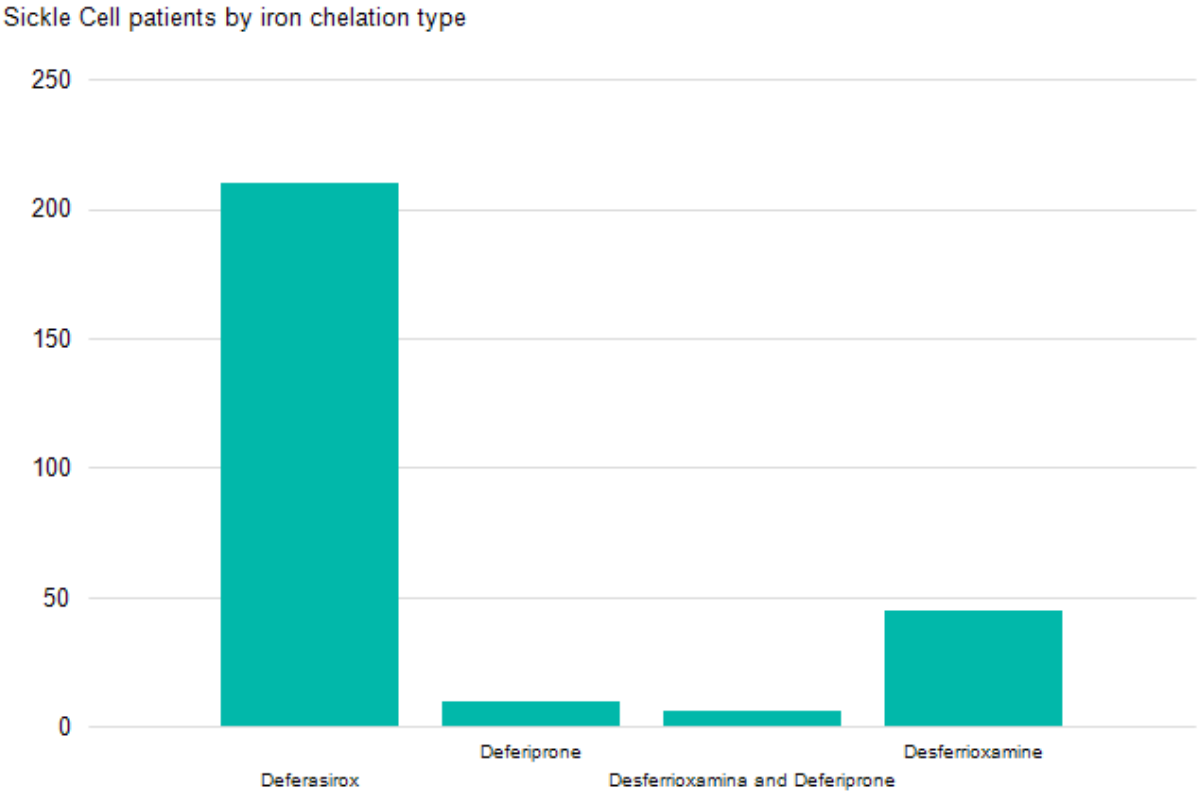


*Figure 5 - Sickle Cell patients by iron chelation type.*

| Treatment Group Id | Treatment | No. |
|---|---|---|
| Chelation Regimen | Deferasirox | 210 |
| | Desferrioxamine | 45 |
| | Deferiprone | 10 |
| | Desferrioxamina and Deferiprone | 6 |
| | Deferasirox and Deferiprone | <5 |
| | Deferasirox and Desferrioxamine | <5 |
| Disease Modifying Therapy | Hydroxycarbamide | 3299 |
| | Prednisolone | 30 |
| | Other | 15 |
| | Dexamethasone | <5 |
| Endocrine | Levothyroxine | 32 |
| | Insulin | 23 |
| | Hydrocortisone | 12 |
| | Oral hypoglycaemic agent | 6 |
| | Ostrogen and progesterone replacement therapy | 5 |
| | Calctriol (rocaltriol) | <5 |
| | Growth hormone | <5 |
| | Testosterone replacement therapy | <5 |
| Other Therapy | Folic acid | 5603 |
| | Penicillin | 5491 |
| | Other | 4896 |
| | Vitamin D | 1746 |
| | Opioid therapy | 246 |
| | Crizanlizumab | 156 |
| | Penicillin alternative | 123 |
| | Ace inhibitor | 85 |
| | Voxelotor | 70 |
| | NOAC/DOAC | 61 |
| | Warfarin | 42 |
| | Etliefrine | 10 |
| | Sildenafil | 8 |
| | Angiotensin receptor blocker | 7 |
| | Home oxygen | 7 |
| | Anti platelet therapy | 6 |
| | Bisphosponate | <5 |
| | Pancreatic enzyme supplement | <5 |
| | Renal replacement therapy | <5 |

*Table 4 - Sickle Cell patient treatments.*

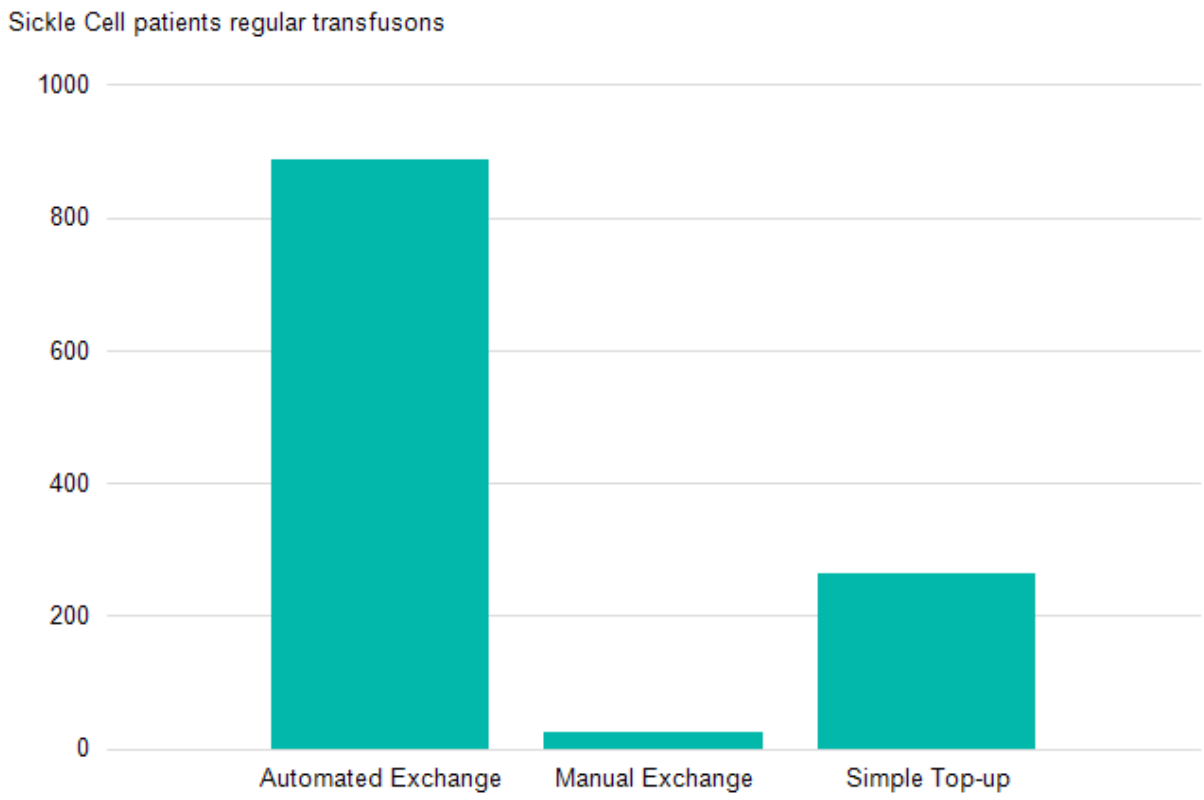


*Figure 6 - Sickle Cell patients receiving regular transfusions by modality.*

| HCC Name | Registrations |
|---|---|
| South East London and South East | 726 |
| East London and Essex | 347 |
| West Midlands | 287 |
| North West | 281 |
| North East and Yorkshire | 264 |
| North Central London and East Anglia | 193 |
| West London | 172 |
| East Midlands | 157 |
| Wessex and Thames Valley | 123 |
| South West | 64 |
| London and South East | <5 |
| No HCC specified | 6 |

*Table 5 - Sickle Cell registrations with TCD monitoring by HCC.*

# Chapter 3: Thalassemia Reports

| SHTName | Registrations |
|---|---|
| Birmingham Women's and Children's Hospital NHS FT and Sandwell and West Birmingham Hospitals NHS Trust | 294 |
| University College London Hospitals NHS Foundation Trust | 240 |
| Whittington Health NHS Trust | 221 |
| Barts Health NHS Trust | 189 |
| Manchester University NHS Foundation Trust | 166 |
| Leeds Teaching Hospitals NHS Trust | 127 |
| Imperial College Healthcare NHS Trust | 90 |
| Oxford University Hospitals NHS Foundation Trust | 74 |
| University Hospitals of Leicester NHS Trust | 69 |
| North Middlesex University Hospital NHS Trust | 66 |
| London Northwest University Healthcare NHS Trust | 64 |
| King's College Hospital NHS Foundation Trust | 50 |
| Nottingham University Hospitals NHS Trust | 48 |
| St Georges Healthcare NHS Foundation Trust | 46 |
| University Hospitals Bristol & Weston NHS Foundation Trust | 46 |
| Guy's and St Thomas' NHS Foundation Trust | 40 |
| The Newcastle Upon Tyne Hospitals NHS Foundation Trust | 34 |
| Royal Liverpool University Hospital (Liverpool University Hospitals NHS Foundation Trust) | 27 |
| Sheffield Teaching Hospitals NHS Foundation Trust | 27 |
| Lewisham and Greenwich NHS Trust | 21 |
| Addenbrooke's Hospital Cambridge (Cambridge University Hospitals NHS Foundation Trust) | 17 |
| University Hospital Southampton NHS Foundation Trust | 15 |
| Sheffield Children's NHS Foundation Trust | 11 |
| Croydon Health Services NHS Trust | 5 |
| Homerton Healthcare NHS Foundation Trust | <5 |
| St James's (Leeds Teaching Hospitals NHS Trust) | <5 |
| University Hospital of Wales (Cardiff and Vale University Health Board) | <5 |
| No SHT specified | 11 |

*Table 6 - Thalassaemia registrations by SHT.*

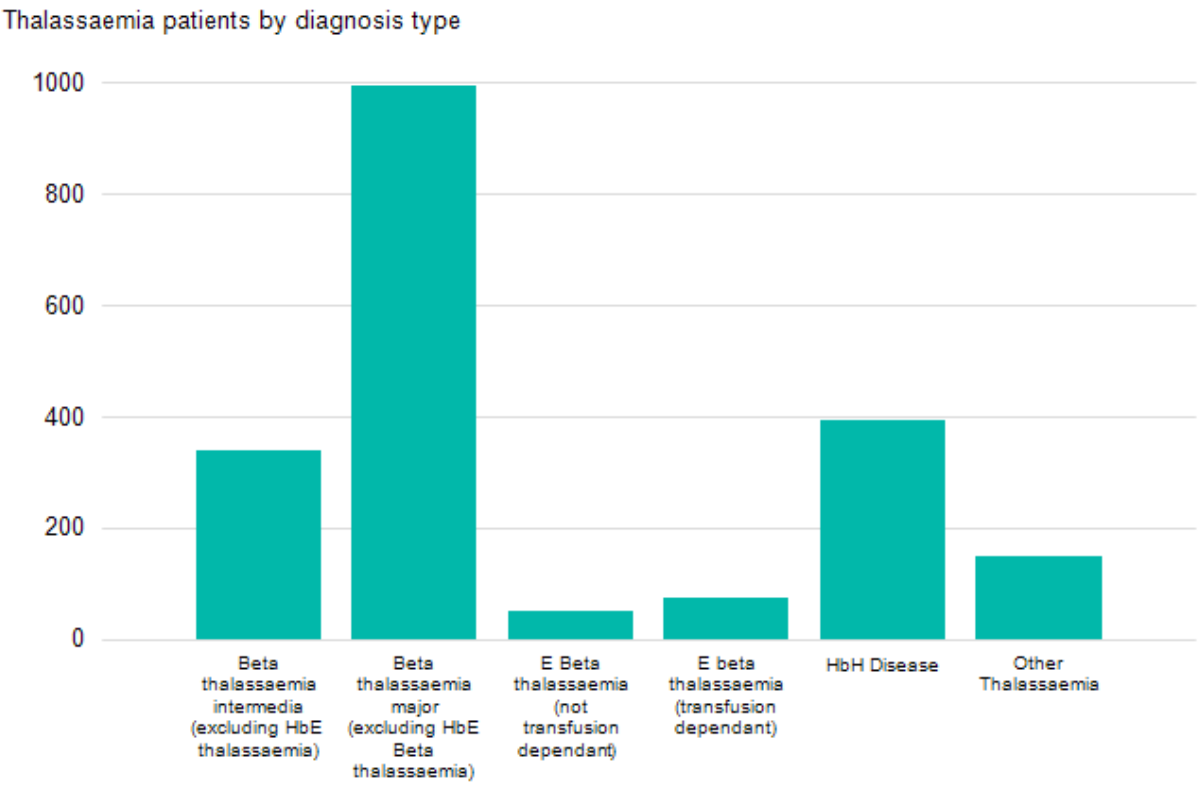


*Figure 7 - Thalassaemia patients by diagnosis type.*

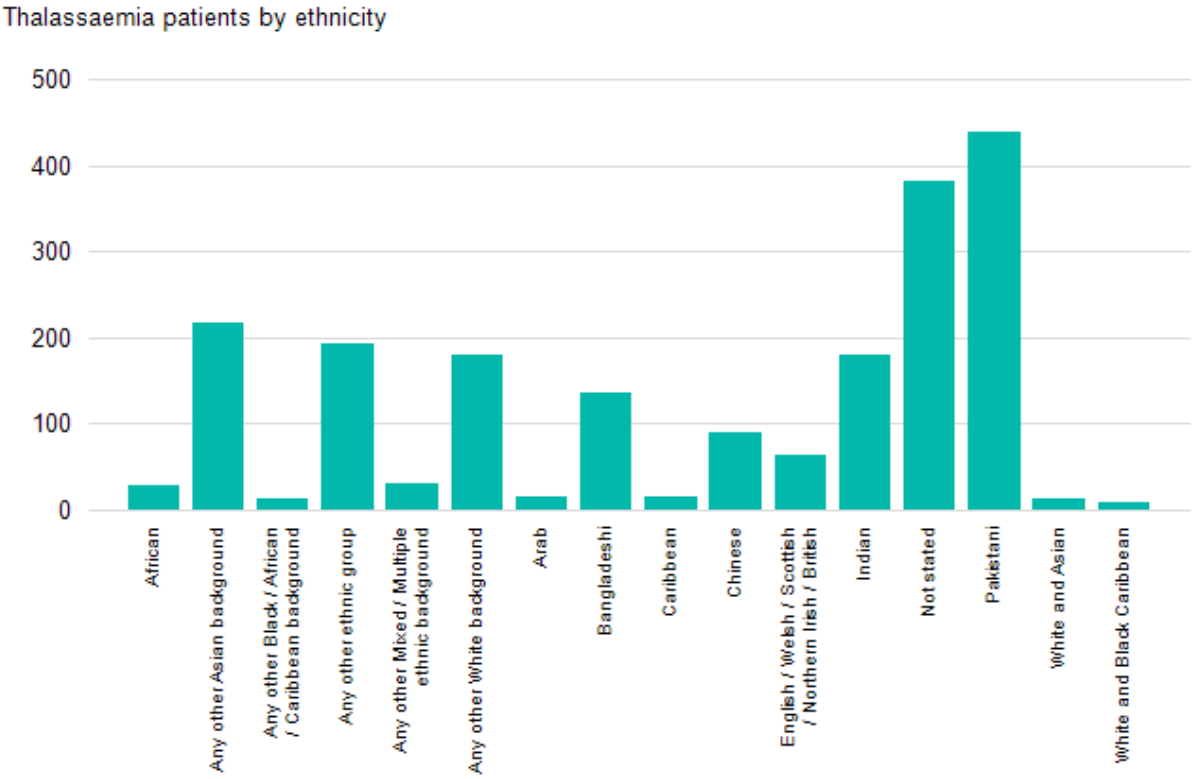


*Figure 8 - Thalassaemia patients by ethnicity.*

| HCC Name | Registrations |
|---|---|
| London, South Central and South West | 878 |
| Midlands | 411 |
| North | 392 |
| London and South East | 305 |
| North Central London and East Anglia | <5 |
| North East and Yorkshire | <5 |
| South East London and South East | <5 |
| No HCC specified | 11 |

*Table 7 - Thalassaemia registrations by HCC.*

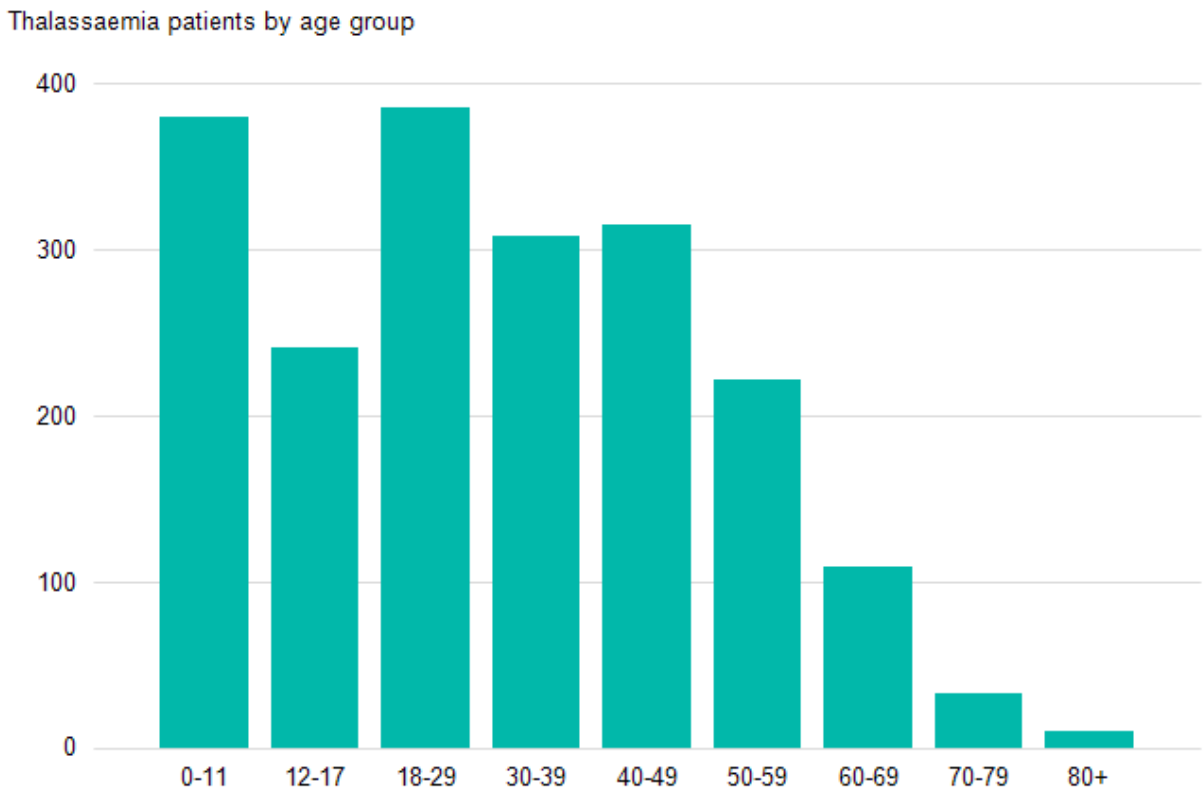


*Figure 9 - Thalassaemia patients by age group.*

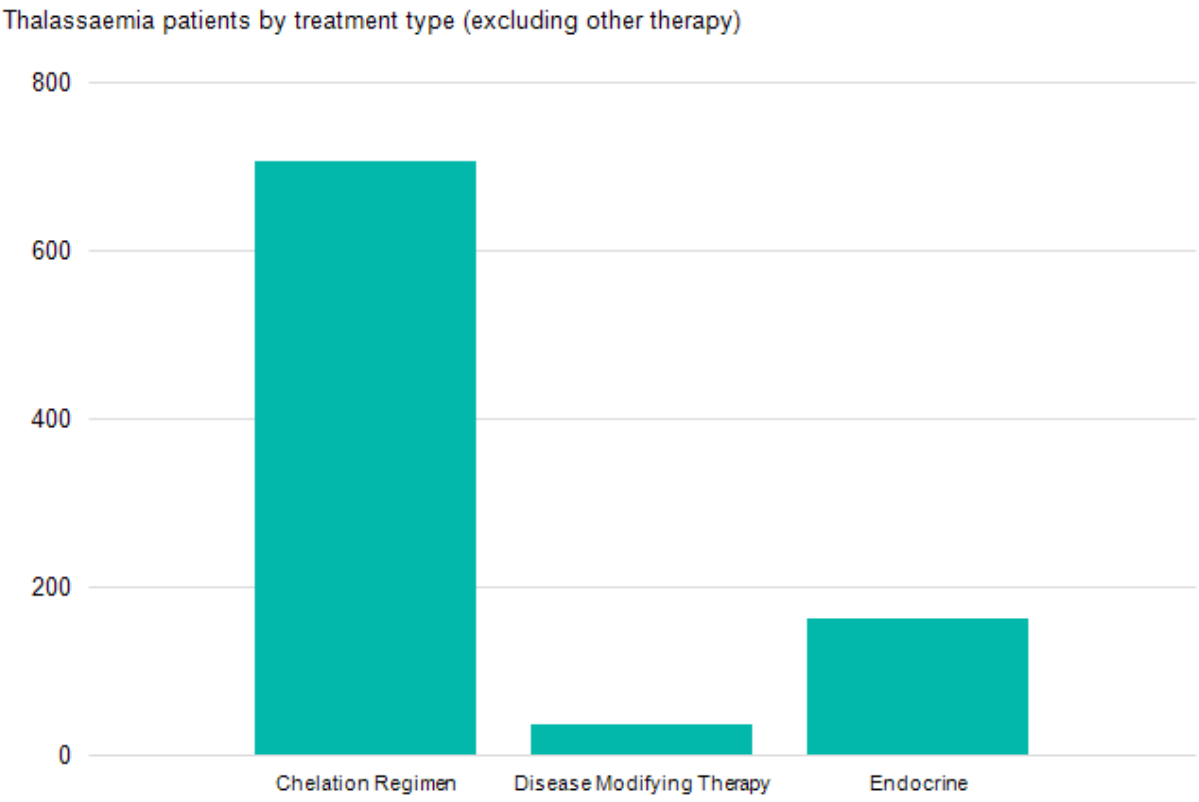


*Figure 10 - Thalassaemia patients by treatment type (excluding other therapy).*

| Treatment group | Treatment | No. |
|---|---|---|
| Chelation Regimen | Deferasirox | 432 |
| | Desferrioxamine | 114 |
| | Deferiprone | 66 |
| | Desferrioxamina and Deferiprone | 34 |
| | Deferasirox and Deferiprone | 32 |
| | Deferasirox and Desferrioxamine | 29 |
| Disease Modifying Therapy | Hydroxycarbamide | 32 |
| | Other | 5 |
| Endocrine | Levothyroxine | 53 |
| | Testosterone replacement therapy | 42 |
| | Insulin | 35 |
| | Ostrogen and progesterone replacement therapy | 15 |
| | Growth hormone | 11 |
| | Calctriol (rocaltriol) | <5 |
| | Fertility therapy | <5 |
| | Hydrocortisone | <5 |
| | Oral hypoglycaemic agent | <5 |
| Other Therapy | Other | 618 |
| | Folic acid | 400 |
| | Vitamin D | 392 |
| | Penicillin | 136 |
| | Warfarin | 15 |
| | Bisphosponate | 10 |
| | NOAC/DOAC | 8 |
| | Sildenafil | 6 |
| | Penicillin  alternative | 5 |
| | Ace inhibitor | <5 |
| | Opioid therapy | <5 |
| | Pancreatic enzyme supplement | <5 |

*Table 8 - Thalassaemia patient treatments.*

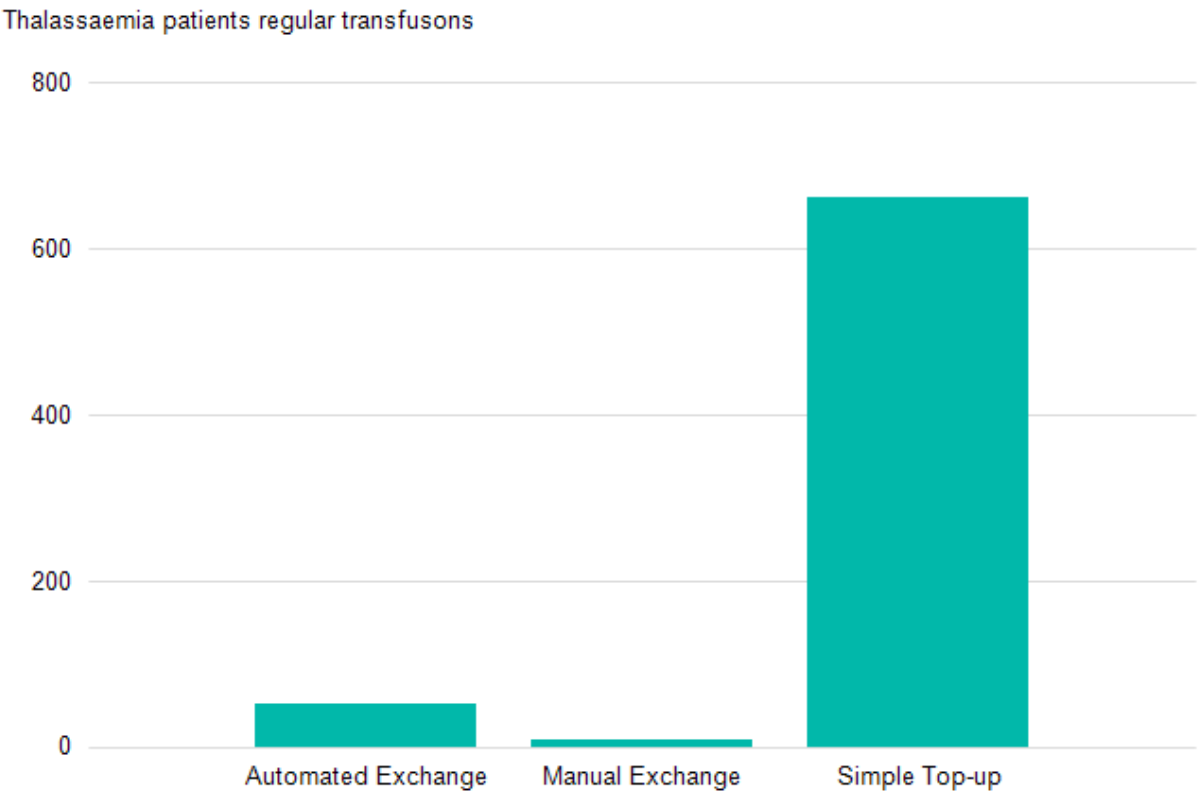


*Figure 11 - Thalassaemia patients regular transfusions by modality.*

# Chapter 4: Rare Inherited Anaemia Reports

| SHT Name | Registrations |
|---|---|
| Birmingham Women's and Children's Hospital NHS FT and Sandwell and West Birmingham Hospitals NHS Trust | 78 |
| Manchester University NHS Foundation Trust | 73 |
| University College London Hospitals NHS Foundation Trust | 63 |
| London Northwest University Healthcare NHS Trust | 52 |
| Oxford University Hospitals NHS Foundation Trust | 36 |
| Nottingham University Hospitals NHS Trust | 31 |
| Barts Health NHS Trust | 26 |
| The Newcastle Upon Tyne Hospitals NHS Foundation Trust | 24 |
| University Hospitals Bristol & Weston NHS Foundation Trust | 20 |
| Royal Liverpool University Hospital (Liverpool University Hospitals NHS Foundation Trust) | 19 |
| Leeds Teaching Hospitals NHS Trust | 17 |
| Sheffield Teaching Hospitals NHS Foundation Trust | 15 |
| University Hospitals of Leicester NHS Trust | 13 |
| Whittington Health NHS Trust | 12 |
| Guy's and St Thomas' NHS Foundation Trust | 9 |
| Homerton Healthcare NHS Foundation Trust | 7 |
| Croydon Health Services NHS Trust | 5 |
| Imperial College Healthcare NHS Trust | 5 |
| King's College Hospital NHS Foundation Trust | 5 |
| University Hospital Southampton NHS Foundation Trust | 5 |
| Addenbrooke's Hospital Cambridge (Cambridge University Hospitals NHS Foundation Trust) | <5 |
| Lewisham and Greenwich NHS Trust | <5 |
| North Middlesex University Hospital NHS Trust | <5 |
| Sheffield Children's NHS Foundation Trust | <5 |
| St Georges Healthcare NHS Foundation Trust | <5 |
| No SHT specified | 72 |

*Table 9 - Rare Inherited Anaemia registrations by SHT.*

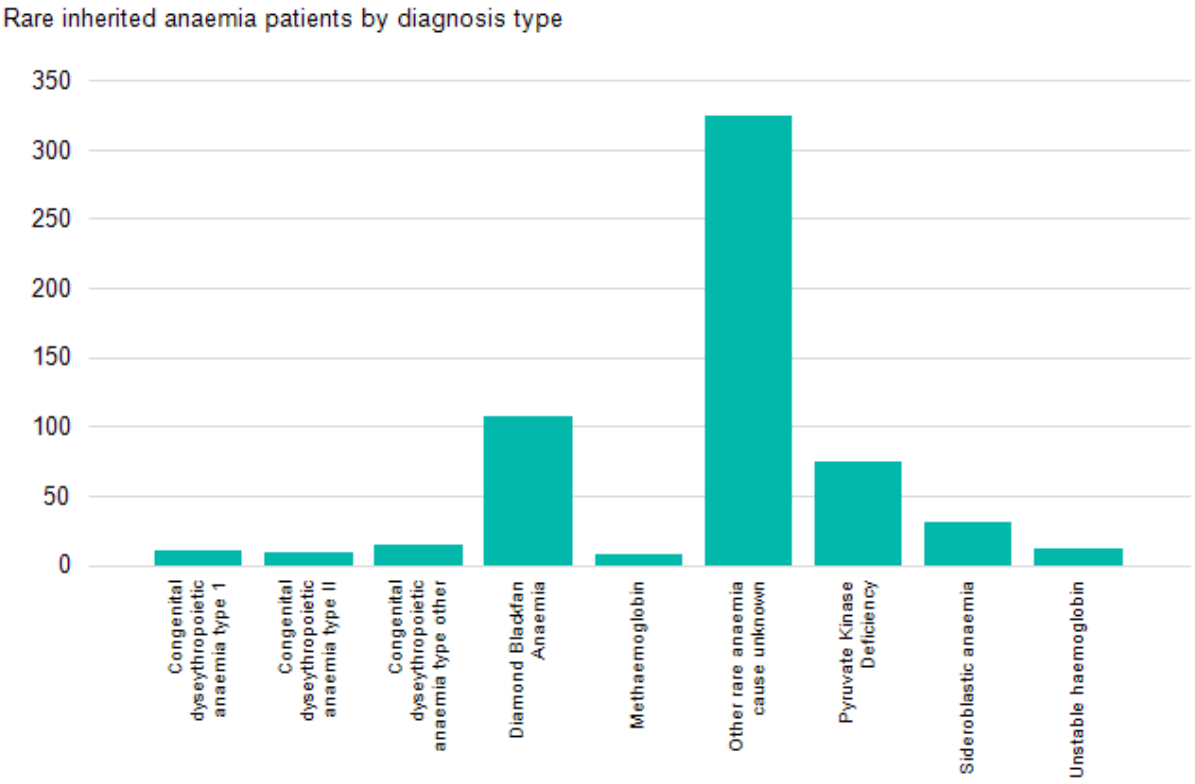


*Figure 12 - Rare Inherited Anaemia patients by diagnosis type.*

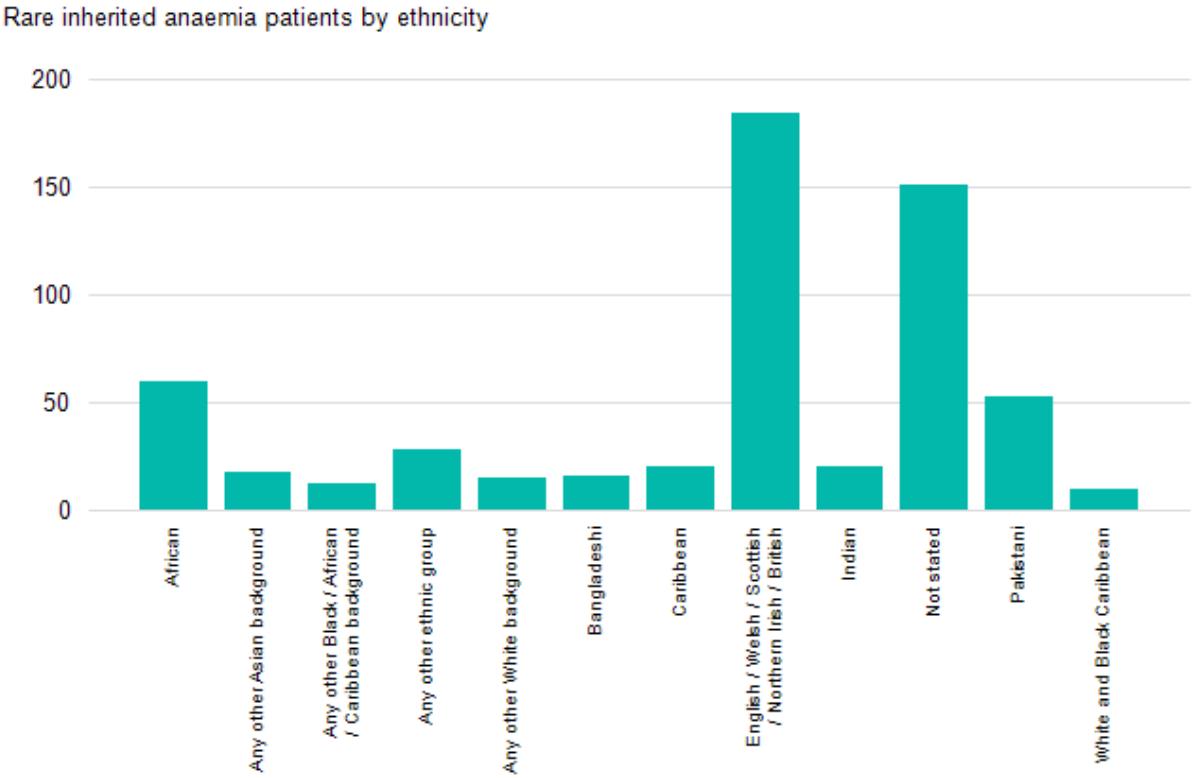


*Figure 13 - Rare Inherited Anaemia patients by ethnicity.*

| HCC Name | Registrations |
|---|---|
| London, South Central and South West | 202 |
| North | 151 |
| Midlands | 122 |
| London and South East | 48 |
| South East London and South East | 6 |
| East London and Essex | <5 |
| North East and Yorkshire | <5 |
| No HCC specified | 70 |

*Table 10 - Rare Inherited Anaemia registrations by HCC.*

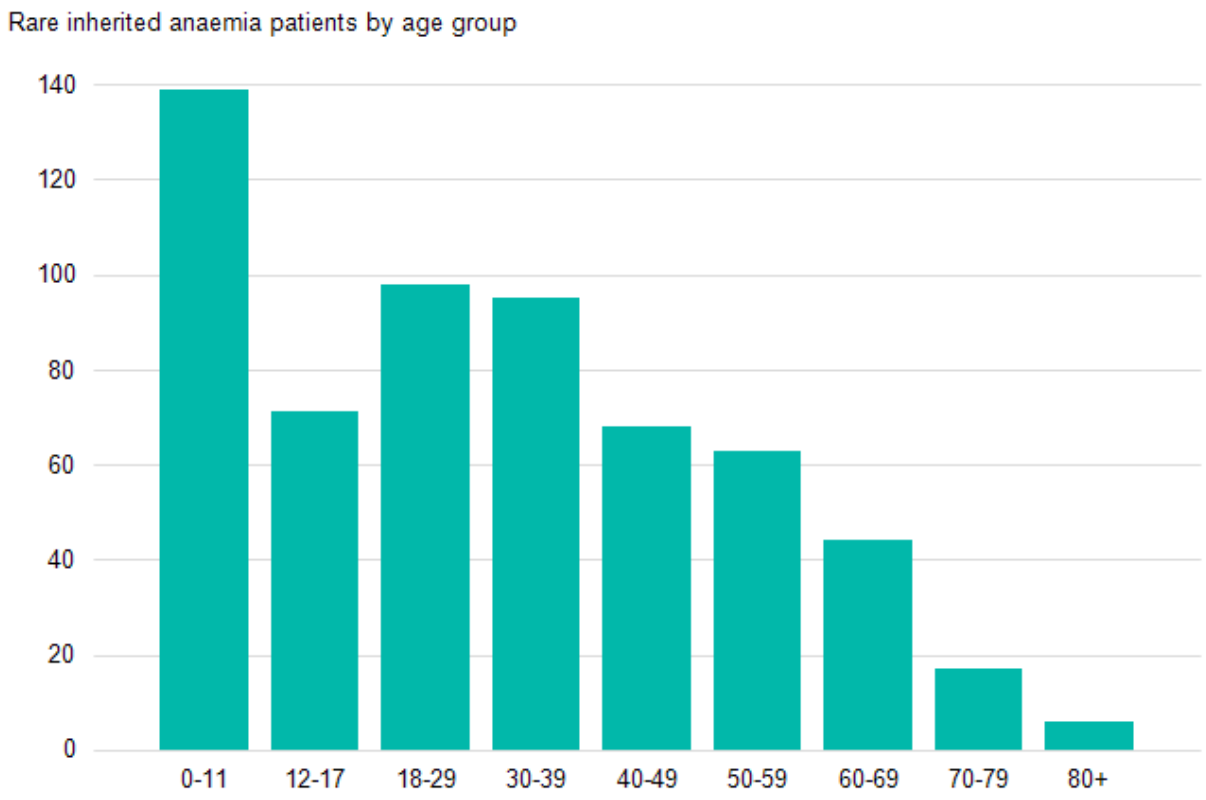


*Figure 14 - Rare Inherited Anaemia patients by age group.*

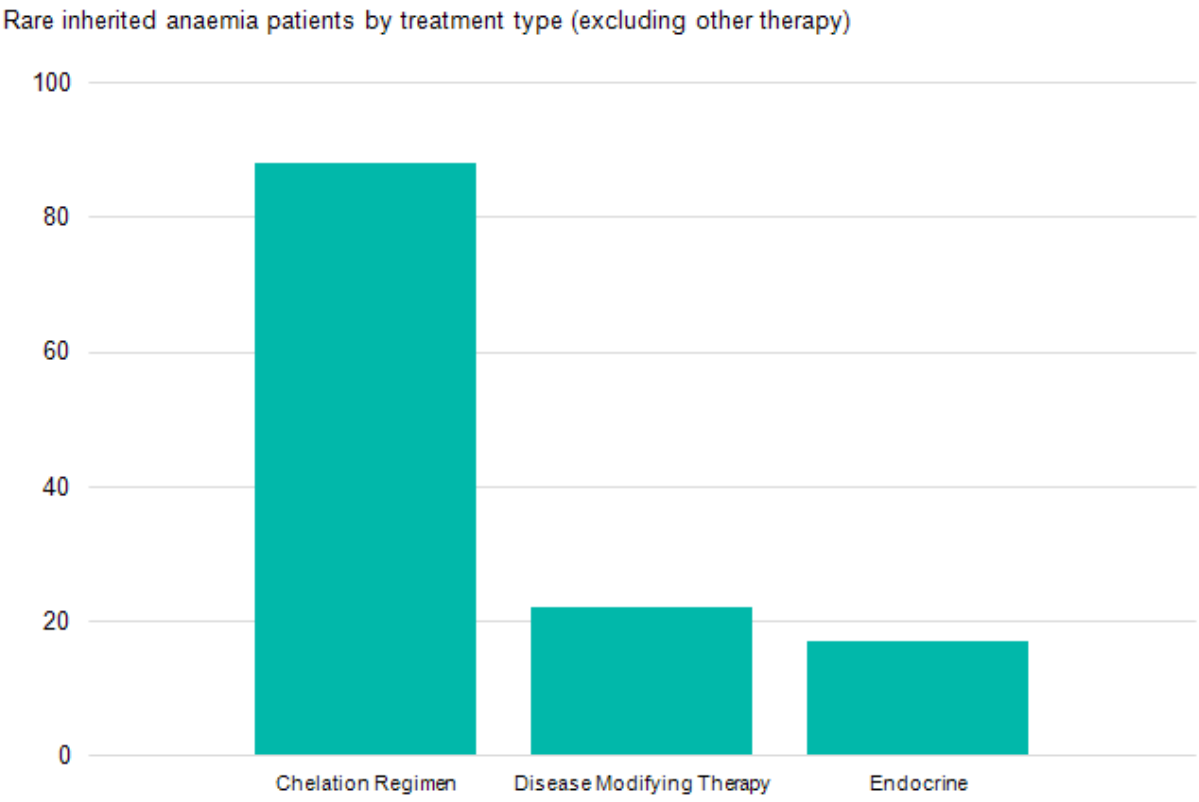


*Figure 15 - Rare Inherited Anaemia patients by treatment type (excluding other therapy).*

| Treatment group | Treatment | No. |
|---|---|---|
| Chelation Regimen | Deferasirox | 61 |
| | Desferrioxamine | 14 |
| | Deferiprone | 5 |
| | Deferasirox and Deferiprone | <5 |
| | Deferasirox and Desferrioxamine | <5 |
| | Desferrioxamina and Deferiprone | <5 |
| Disease Modifying Therapy | Prednisolone | 10 |
| | Hydroxycarbamide | 9 |
| | Interferon | <5 |
| Endocrine | Levothyroxine | 9 |
| | Growth hormone | <5 |
| | Insulin | <5 |
| | Testosterone replacement therapy | <5 |
| Other Therapy | Other | 114 |
| | Folic acid | 107 |
| | Vitamin D | 60 |
| | Penicillin | 38 |
| | Bisphosponate | <5 |
| | Home oxygen | <5 |
| | NOAC/DOAC | <5 |
| | Penicillin alternative | <5 |
| | Warfarin | <5 |

*Table 11 - Rare Inherited Anaemia patient treatments.*

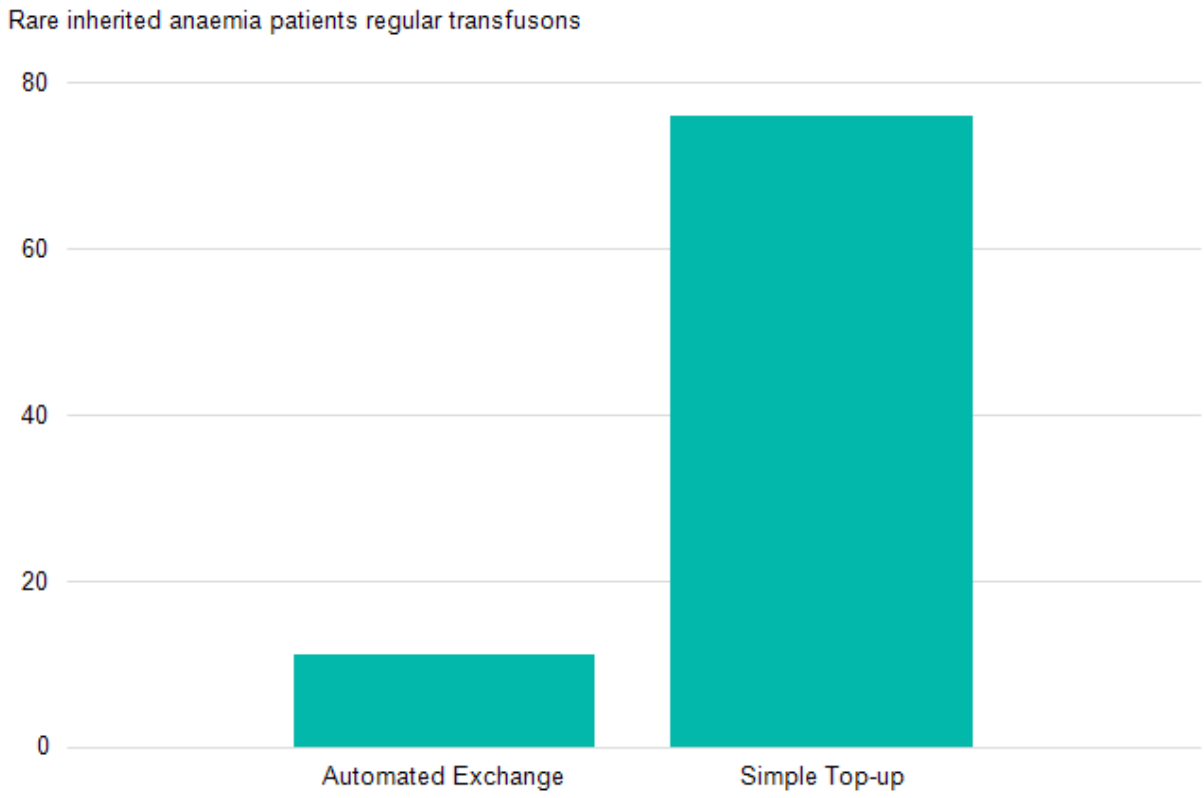


*Figure 16 - Rare Inherited Anaemia patients regular transfusions by modality.*

# Chapter 5: Reported Serious Events and Comorbidities.

| Serious adverse events | No. |
|---|---|
| Death | 30 |
| Delayed HTR - Not thought to be associated with antibody: hyperhaemolysis | 10 |
| Ischaemic stroke | 8 |
| Cardiac failure | 6 |
| Delayed HTR - Associated with a new antibody (conventional DHTR) | <5 |
| Delayed HTR - Associated with previous antibody | <5 |
| Haemorrhagic stroke | <5 |
| Intrauterine death | <5 |
| Pneumococcal Infection | <5 |

*Table 12 - Sickle Cell Serious Adverse Events.*

| Comorbidity type | Comorbidity | No. |
|---|---|---|
| Acute Haemolytic Transfusion Reaction (HTR Acute) | Acute HTR - Not thought to be associated with antibody: hyperhaemolysis | 9 |
| | Acute HTR - Associated with a new antibody (conventional DHTR) | <5 |
| Bacterial infection | Bacterial infectious disease (Other) | 20 |
| Cardiorespiratory | Acute chest syndrome | 114 |
| | Pneumonia | 16 |
| | Pulmonary embolism | 6 |
| | Deep vein thrombosis | 5 |
| | Fat embolism syndrome | 5 |
| | Asthma | <5 |
| | Obstructive lung disease | <5 |
| | Pulmonary hypertension | <5 |
| Endocrine | Other endocrine complication | <5 |
| Genitourinary | Priapism | 15 |
| | Acute renal failure | <5 |
| | Chronic renal failure Stage 3 CKD: eGFR Between 30 and 59 | <5 |
| | Chronic renal failure Stage 4 CKD: eGFR Between 15 and 29 | <5 |
| | Papillary necrosis | <5 |
| Haematological | Simple painful crisis | 852 |
| | Acute haemolyitc event not related to blood transfusion | <5 |

| | | |
|---|---|---|
| Hepatobiliary | Acute Pancreatitis | <5 |
| | Ascending cholangitis | <5 |
| | Cirrhosis of liver | <5 |
| | Hepatic sequestration | <5 |
| | Liver failure | <5 |
| Neurological disorders | Chronic pain | 12 |
| | Epilepsy | <5 |
| | Retinopathy Stage I Peripheral aterial occlusion | <5 |
| | Retinopathy Stage II Peripheral arteriovenous anastomoses (hairpin loop) | <5 |
| | Retinopathy Stage III Neovascular and fibrous proliferations (sea fan) | <5 |
| | Retinopathy Stage IV Vitreous hemorrhage | <5 |
| | Seizure without diagnosis of epilepsy | <5 |
| | Silent cerebral infarcts | <5 |
| | Visual loss | <5 |
| Obstetric/ gynaecological | Recurrent miscarrage | <5 |
| Orthopaedic | Avascular necrosis | 18 |
| | Leg ulcers | 12 |
| | Acute Osteomyelitis | 10 |
| | Chronic Osteomyelitis | <5 |
| | Fracture | <5 |
| | Osteopenia | <5 |
| Other | Other complication not listed above | 117 |
| | Splenic sequestration | 14 |
| | Cancer | <5 |
| Viral infection | Covid 19 | 88 |
| | Influenza | 19 |
| | Other viral illness | 14 |
| | Heptitis B | <5 |
| | Parvovirus | <5 |

*Table 13 - Sickle Cell comorbidities.*

| Serious adverse events | No. |
|---|---|
| Cardiac failure | <5 |
| Delayed HTR - Not thought to be associated with antibody: hyperhaemolysis | <5 |

*Table 14 – Thalassaemia patients serious adverse events.*

| Comorbidity type | Comorbidity | No. |
|---|---|---|
| Bacterial infection | Bacterial infectious disease (Other) | <5 |
| Cardiorespiratory | Deep vein thrombosis | <5 |
| | Pneumonia | <5 |
| Endocrine | Insulin dependant diabetes | <5 |
| Genitourinary | Hydronephrosis | <5 |
| | Renal replacement therapy | <5 |
| | Renal stones | <5 |
| Haematological | Simple painful crisis | <5 |
| Orthopaedic | Fracture | <5 |
| | Osteoporosis | <5 |
| Other | Other complication not listed above | 16 |
| | Cancer | <5 |
| Viral infection | Covid 19 | 9 |
| | Influenza | <5 |
| | Other viral illness | <5 |
| | Parvovirus | <5 |

*Table 15 - Thalassaemia patients comorbidities.*

| Serious adverse events | No. |
|---|---|
| Cardiac failure | <5 |
| Death | <5 |

*Table 16 - Rare Inherited Anaemia Serious Adverse Events*

| Comorbidity type | Comorbidity | No. |
|---|---|---|
| Bacterial infection | Bacterial infectious disease (Other) | <5 |
| Cardiorespiratory | Pneumonia | <5 |
| Endocrine | Adrenal insufficicency | <5 |
| Haematological | Neutropenia | <5 |
| | Simple painful crisis | <5 |
| Other | Other complication not listed above | <5 |
| Viral infection | Covid 19 | <5 |
| | Influenza | <5 |

*Table 17 - Rare Inherited Anaemia Comorbidities*