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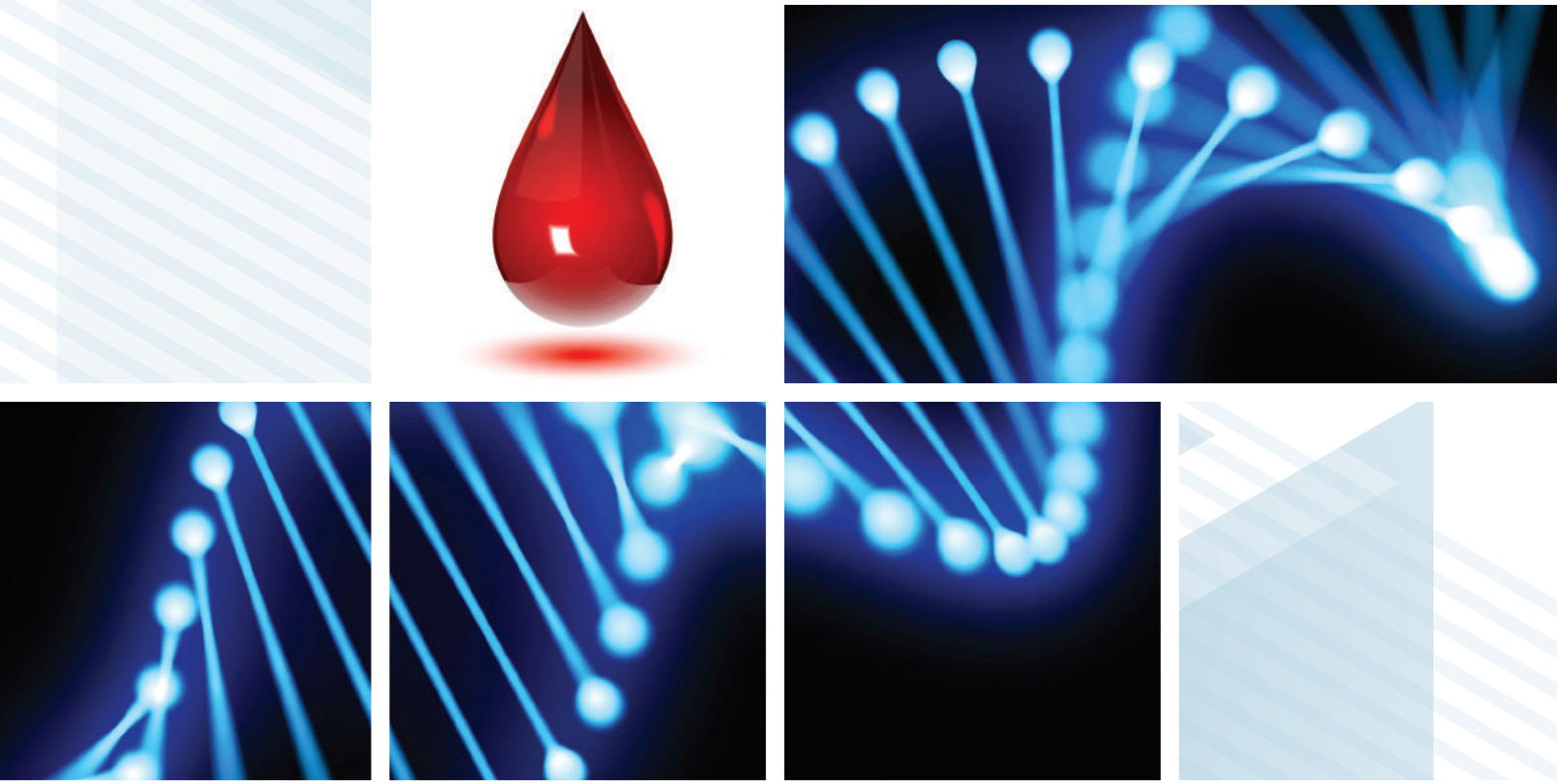
ANNUAL REPORT OF THE MALAYSIAN THALASSAEMIA REGISTRY 2019



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Ministry of Health Malaysia

FOREWORD FROM THE MINISTER OF HEALTH MALAYSIA

Thalassaemia is a chronic debilitating disease affecting nearly 200 million people worldwide and has a significant impact on the community and the Malaysian health system. Reducing the incidence of thalassaemia, while ensuring the best quality care and outcomes for those who experience the disease, are key priorities taken by Ministry of Health and other related agencies overall.

The Malaysian Thalassaemia Registry (MTR) launched on the 12th of May 2007 is part of the National Thalassaemia Prevention and Control Programmes. Since 2007, MTR has led the reporting of thalassaemia scenario in the country based on detailed epidemiological and clinical data collected through 13 states and 3 federal territories with total of 110 participating treatment centres. MTR is the largest thalassaemia registry in the world with 8,171 thalassaemia patients to date.

These data enable our health system to identify opportunities to improve thalassaemia prevention initiatives; enhance screening programs; and optimise treatment services and clinical trials and research at a national and local level. Early detection, screening, and diagnosis have been proven significantly to improve patients' survival rates and quality of life as well as significantly reduce the cost and complexity of thalassaemia treatment. However, barriers to achieving higher standard of care need to be addressed now at the individual, health system, and governmental level to significantly reduce the personal and financial burden of thalassaemia worldwide.

Finally, I would like to express my gratitude and congratulate the editorial team for their efforts in producing this important report. We look forward to the continuation of such efforts in future publications and provide key data and information across the full spectrum of thalassaemia multidisciplinary care, including a greater number of indicators that provides a more comprehensive view of thalassaemia health system performance.

Dato' Sri Dr Adham bin Baba
Minister of Health, Malaysia

FOREWORD FROM THE DIRECTOR GENERAL OF HEALTH

I would like to acknowledge the exceptional teamwork by National Thalassaemia Registry team members who have been involved in gathering, processing and producing the data for the Malaysian Thalassaemia Registry 2019. This is the second thalassaemia registry report published by Malaysian Thalassaemia Registry.

To understand the impact thalassaemia has in our country, it is important to monitor the number of people who are affected by the disease, and to what extent. For this reason, the Thalassaemia Registry regularly gathers information about the number of people diagnosed with thalassaemia, the different type of thalassaemia, what treatment have they received, the complications of iron overload and the mortality. It is important to look at these data over time. This will show any changes, and look at whether there are any differences between genders, ethnicity groups and geographical areas.

The data from this registry will enhance in tailoring the national thalassaemia programmes, harmonisation of pertinent allotment of resources and dissemination of thalassaemia comprehensive care at a national, state and local level. The success of thalassaemia care is reflected by early detection, prompt treatment and survival improvement after the patient being diagnosed and reducing trend in new birth thalassaemia.

I wish to commend the efforts and sincere appreciation and gratitude to all clinicians, pathologists, pharmacists, radiologists, research assistants for their contribution to this report. This report marked another milestone in strengthening thalassaemia management in this country.

Tan Sri Dr Noor Hisham bin Abdullah
Director General of Health, Malaysia

EXECUTIVE SUMMARY

The Malaysian Thalassaemia Registry (MTR) was launched on the 12th of May 2007 and its first report was released in May 2019, in conjunction with the World Thalassaemia Day. Since the launch, the MTR has achieved many important milestones including maturity of its data. The first MTR Report has been recently published (Ibrahim HM et al., 2020). It is our pleasure to again share with you the latest MTR report for 2019. This report is a collective effort of all personnel who were involved in thalassaemia care across the country to prospectively capture the real world clinical experience. The substantial positive impact of the government involvement in the National Thalassaemia Programme, which includes screening programme, public awareness, accurate diagnosis, accessibility to iron chelators, improvement of blood transfusions service and disease monitoring facilities, is reflected in this second report. The information gathered is important to facilitate the health division strategies for optimal resources distribution among the states.

The year 2019 report saw 194 new patients included into the registry, making up a total of 8178 patients. The rising number of total patients was contributed by increased survival rates, better reporting of newly diagnosed patients by capture of the less severe forms of thalassaemia, and improved accessibility and compliance to iron chelators. Overall, the trend of yearly new cases especially from 2015 onwards is declining in numbers, which reflects the positive return on the government investment for the National Thalassaemia Programme. MTR has successfully mapped the geographical distribution of thalassaemia diagnosis among the Malaysian multi-ethnic population. It is observed that β-thalassaemia major is the most common diagnosis in Sabah, predominantly in the Kadazan-Dusun, while in Peninsular Malaysia HbE/β-thalassaemia is the most common diagnosis, with majority of Malay patients. A high proportion of Chinese patients were diagnosed with Haemoglobin H (HbH) disease in comparison to other ethnicities, with a life expectancy of beyond 55 years old.

Improvement in the management of iron overload and improved compliance to iron chelators especially the oral preparations most likely explain for the declined levels of serum ferritin observed in the past few years. However, the mortality rates among the thalassaemics were between 6.2-8.6 per 1000 patients per year in the past 5 years. The main cause of death was cardiac failure. These findings emphasise the importance of more effective iron chelation therapy, good compliance and a dire need for end organ iron overload monitoring, especially magnetic resonance imaging (MRI) T2* and not only serum ferritin levels.

Moving forward, we hope that the MTR will soon incorporate the i) molecular diagnosis, for an early, complete and accurate diagnosis, ii) classification of patients into transfusion-dependent thalassaemia (TDT) or non-transfusion-dependent thalassaemia (NTDT), iii) MRI T2* results, iv) other end organ complications (particularly endocrine complications) and v) detailed causes of death (including the organisms involved in deaths due to sepsis). This will enable local health authorities and health providers to plan cost-effective services, reduce healthcare burden, reduce mortality and ultimately improve the quality of life of all thalassaemics. A recent study on health-related quality of life among Malaysian children with TDT showed improvement over the last decade owing to the better access in treatment (Shafie AA et al., 2020).

Sustainability of the MTR is important. Therefore, we urge the government, policymakers and research funding bodies to continuously support the registry for optimal care of local thalassaemics. We trust the database will serve as a robust tool for supporting research and advocacy initiatives, and pushing boundaries of care for thalassaemics for many years to come.

Editors

ACKNOWLEDGEMENTS

We would like to thank and acknowledge the contribution of the following:

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1 INTRODUCTION

1.1 Thalassaemia

Inherited disorders of haemoglobin are the most common monogenic diseases worldwide. Thalassaemia is one of the most common autosomal recessive disorders involving abnormal haemoglobin formation with high frequency in tropical countries including Malaysia. It resulted from reduced production of one or more sub-units of haemoglobin which comprises both α- and β-globin chains. The two main categories of thalassaemia are α-thalassaemia and β-thalassaemia; these are further divided into several subcategories. β-thalassaemia major causes severe anaemia which requires life-long blood transfusion for survival. The World Health Organisation has indicated thalassaemia as a major public health concern, and accurate information regarding its health burden in countries with high prevalence of thalassaemia is needed. Although thalassaemia is the most common hereditary haematological disorder in Malaysia, information on geographical distribution of patients, socioeconomic data and clinical data including treatment outcomes is still lacking. It is estimated that about 4.5% of the Malaysians are carriers (George, 1998). The molecular defects which lead to β-thalassaemia are heterogeneous, and each ethnic group possesses its own specific set of mutations. β-thalassaemia is more prominent among the Malay and Kadazan-Dusun, while it is rarely found among the Malaysian Indian. Haemoglobin (Hb) Bart's hydrops fetalis has been reported mainly among the Malaysian Chinese (Wee Y-C et al., 2005).

In transfusion-dependent thalassaemia (TDT) patients, regular blood transfusion is required as it alleviates symptomatic anaemia, suppresses ineffective erythropoiesis, reduces iron loading from increased gastrointestinal absorption, enhances growth and development, and improves survival rate. The transfusion goal is to maintain a pre-transfusion haemoglobin level of between 9 and 10 g/dL (Chonat and Quinn, 2018). For non-transfusion-dependent thalassaemia (NTDT) patients, they might require blood transfusion if they experience symptomatic anaemia, poor quality of life, compromised growth and development, or if there are features of extramedullary haematopoiesis. Patients with transfusional iron overload will require iron chelation therapy to help decrease the iron burden and to prevent long-term complications associated with end organ iron deposition in tissues, such as hepatic dysfunction and failure, endocrinopathies, and cardiac dysfunction.

Due to concern over its public health burden on the country, the Cabinet of Malaysia endorsed a national comprehensive thalassaemia programme on 25 August 2004, consisting of health education and population awareness drive, screening initiative, comprehensive clinical management and establishing a Malaysian Thalassaemia Registry (MTR). The MTR was first started in IPHKL and PPUKM in 2006. Initially, data was collected only for IPHKL and PPUKM using Microsoft Access, after which, the data was migrated to the current website-based registry format. Data entry training for end users commenced in 2007. The registry was officially launched on the 12th of May 2007 with the objective to identify and collect detailed epidemiological and clinical data of patients with thalassaemia from all over the country, who received treatment at government hospitals under the Ministry of Health (MOH) or university hospitals under the Ministry of Education. The registry is the first online patient registry in Malaysia featuring real-time data entry, which facilitates update and data reporting, and allows enrolled users to observe the aggregated data at any point of time. The essential data elements were predefined by a team of experts including clinicians, and the completeness and validity of data collection was ensured by joint cooperation between the company in-charge and MOH, Malaysia. Site visits to ensure data accuracy and completeness were carried out by a team of research assistants appointed by the MOH and working in collaboration with the company. A regular internal audit for quality control of the MTR is performed by the company in-charge. The web-based system is accessible to enrolled users through www.mytalasemia.net.my (MyTalasemia). The system is user-friendly and can be managed from different locations. The MTR demonstrates the value of a continuously updated registry for the surveillance of health services pertaining to thalassaemia in the country. Patient registries, which include a large set of data, have been reported to be helpful in mapping the functionalities and providing a positive return on investment.

1.2 Demographics in Malaysia

Malaysia consists of 13 states and 3 federal territories. Eleven states and 2 federal territories are located in the Peninsular Malaysia. The other 2 states are on the island of Borneo, and the remaining one federal territory consists of an island offshore of Borneo; these are collectively referred to as East Malaysia. The list of states and federal territories of Malaysia are as follows:

1. Johor
2. Kedah
3 Kelantan
4. Melaka
5. Negeri Sembilan
6. Pahang
7. Perak
8. Perlis
9. Pulau Pinang
10. Sabah
11. Sarawak
12. Selangor
13. Terengganu
14. W.P. Kuala Lumpur
15. W.P. Labuan
16. W.P. Putrajaya

1.3 Population of Malaysia

Malaysia is a fast-developing country in Southeast Asia, with a population of 32.68 million in fourth quarter of 2019. The citizens are multi-ethnic with a majority of Malay (63.1%), followed by Chinese and Indian ethnicities. Non-citizens make up 8.2% of the population (Department of Statistics Malaysia, 2010).

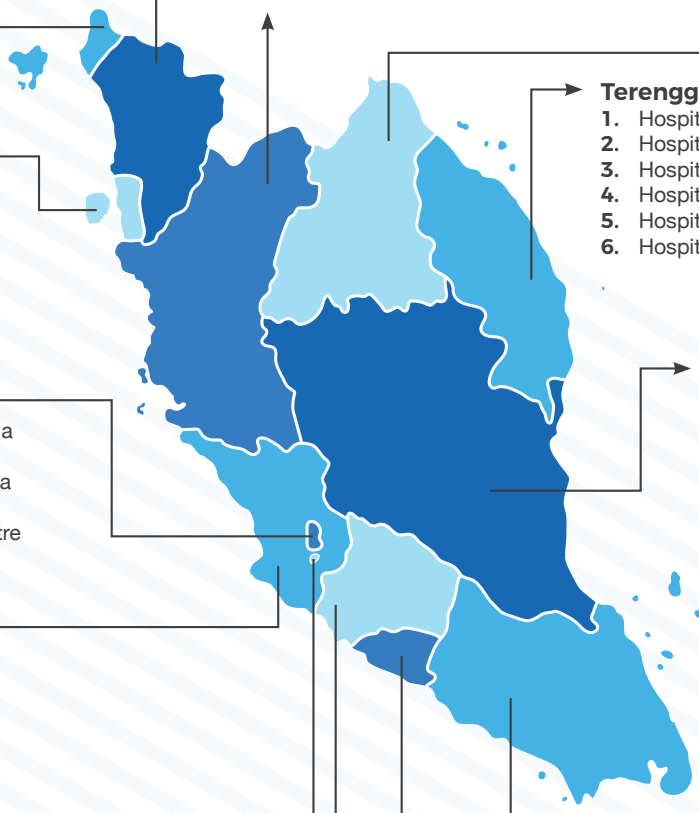
The Malay are the original inhabitants of peninsular Malaysia and together with the indigenous peoples in Sabah and Sarawak, they are known as “Bumiputera” and form the largest community in Malaysia, at 62% of total population. The Malaysian Chinese are descendants of ancestors from South China, whereas the Malaysian Indian are descended from ancestors from Southern India. Aboriginal people known as Orang Asli are present in the East and Peninsular Malaysia.

1.4 Data Collection

The updated MTR includes all patients diagnosed with thalassaemia from 2007 to October 2019. Updated data from the registry was retrieved in November 2019 for analysis. The patients’ data from diagnosis to last follow-up or death were collected by research assistants in various regional centres in Malaysia. All data were verified by clinicians before being manually entered into the MyTalasemia system. All research assistants had undergone central training on data collection and recording into the web-based system. The types of data collected were guided by the registry design. Data elements were grouped into several categories including socio-demography, clinical characteristics, laboratory test results, type of treatment received, death record and complications. Duplicate registration was prevented. The updated data were further verified and discussed centrally during the National Thalassaemia Meeting held annually in November.

1.5 Participating Treatment Centres

West Malaysia



Kedah
1. Hospital Sultanah Bahiyah, Alor Setar
2. Hospital Kulim
3. Hospital Sultan Abdul Halim Shah, Sungai Petani
4. Hospital Baling
5. Hospital Jitra
6. Hospital Kuala Nerang
7. Hospital Langkawi
8. Hospital Sik
9. Hospital Yan

Perlis
1. Hospital Tuanku Fauziah, Kangar

Pulau Pinang
1. Hospital Pulau Pinang
2. Hospital Seberang Jaya
3. Hospital Balik Pulau
4. Hospital Bukit Mertajam
5. Hospital Kepala Batas
6. Hospital Sungai Bakap

W.P. Kuala Lumpur
1. Institut Pediatrik, Hospital Kuala Lumpur
2. Universiti Kebangsaan Malaysia Medical Centre
3. Universiti Malaya Medical Centre
4. Institut Perubatan Respiratori

Selangor
1. Hospital Ampang
2. Hospital Banting
3. Hospital Kajang
4. Hospital Kuala Kubu Bharu
5. Hospital Selayang
6. Hospital Serdang
7. Hospital Sungai Buloh
8. Hospital Tanjung Karang
9. Hospital Tengku Ampuan Jemaah, Sabak Bernam
10. Hospital Tengku Ampuan Rahimah, Klang
11. Hospital Shah Alam

W.P. Putrajaya
1. Hospital Putrajaya

Negeri Sembilan
1. Hospital Tuanku Ja'afar Seremban
2. Hospital Tuanku Ampuan Najihah, Kuala Pilah
3. Hospital Jelebu
4. Hospital Port Dickson
5. Hospital Tampin

Melaka
1. Hospital Melaka
2. Hospital Alor Gajah
3. Hospital Jasin

Perak
1. Hospital Kuala Kangsar
2. Hospital Seri Manjung
3. Hospital Slim River
4. Hospital Taiping
5. Hospital Teluk Intan
6. Hospital Bahagia
7. Hospital Batu Gajah
8. Hospital Raja Permaisuri Bainun
9. Hospital Changkat Melintang
10. Hospital Kampar
11. Hospital Parit Buntar
12. Hospital Selama
13. Hospital Sungai Siput
14. Hospital Tapah
15. Hospital Grik

Kelantan
1. Hospital Gua Musang
2. Hospital Raja Permaisuri Zainab II, Kota Bharu
3. Hospital Kuala Krai
4. Hospital Machang
5. Hospital Pasir Mas
6. Hospital Tanah Merah
7. Hospital Tengku Anis, Pasir Puteh
8. Hospital Tumpat
9. Hospital Universiti Sains Malaysia, Kubang Kerian
10. Hospital Jeli

Terengganu
1. Hospital Besut
2. Hospital Dungun
3. Hospital Hulu Terengganu
4. Hospital Kemaman
5. Hospital Sultanah Nur Zahirah
6. Hospital Setiu

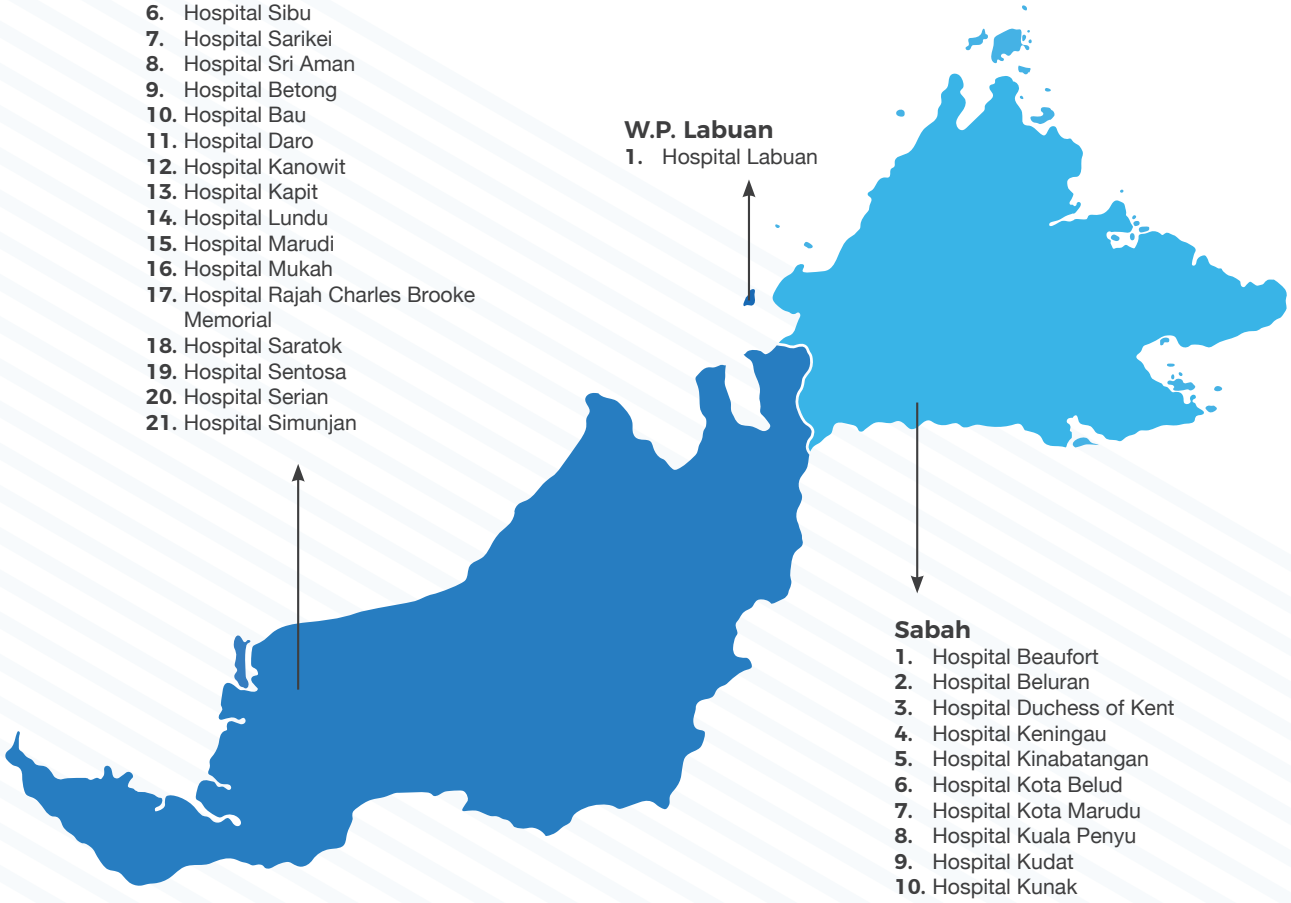
Pahang
1. Hospital Bentong
2. Hospital Jengka
3. Hospital Jerantut
4. Hospital Mentakab
5. Hospital Kuala Lipis
6. Hospital Muadzam Shah
7. Hospital Raub
8. Hospital Sultan Haji Ahmad Shah, Temerloh
9. Hospital Tengku Ampuan Afzan, Kuantan
10. Hospital Pekan
11. Hospital Hajjah Kalsom, Cameron Highlands

Johor
1. Hospital Sultanah Nora Ismail, Batu Pahat
2. Hospital Sultan Ismail, Johor Bharu
3. Hospital Enche' Besar Hajjah Khalsom, Kluang
4. Hospital Kota Tinggi
5. Hospital Mersing
6. Hospital Pakar Sultanah Fatimah, Muar
7. Hospital Pontian
8. Hospital Segamat
9. Hospital Sultanah Aminah, Johor Bharu
10. Hospital Temenggung Seri Maharaja Tun Ibrahim, Kulai
11. Hospital Tangkak

East Malaysia

Sarawak
1. Hospital Bintulu
2. Hospital Umum Sarawak, Kuching
3. Hospital Lawas
4. Hospital Limbang
5. Hospital Miri
6. Hospital Sibu
7. Hospital Sarikei
8. Hospital Sri Aman
9. Hospital Betong
10. Hospital Bau
11. Hospital Daro
12. Hospital Kanowit
13. Hospital Kapit
14. Hospital Lundu
15. Hospital Marudi
16. Hospital Mukah
17. Hospital Rajah Charles Brooke Memorial
18. Hospital Saratok
19. Hospital Sentosa
20. Hospital Serian
21. Hospital Simunjan

W.P. Labuan
1. Hospital Labuan



Sabah
1. Hospital Beaufort
2. Hospital Beluran
3. Hospital Duchess of Kent
4. Hospital Keningau
5. Hospital Kinabatangan
6. Hospital Kota Belud
7. Hospital Kota Marudu
8. Hospital Kuala Penyu
9. Hospital Kudat
10. Hospital Kunak
11. Hospital Lahad Datu
12. Hospital Wanita & Kanak-Kanak Sabah, Likas
13. Hospital Papar
14. Hospital Pitas
15. Hospital Queen Elizabeth, Kota Kinabalu
16. Hospital Ranau
17. Hospital Semporna
18. Hospital Sipitang
19. Hospital Tambunan
20. Hospital Tawau
21. Hospital Tenom
22. Hospital Tuaran

2 REGISTRY REPORT

There are 137 government hospitals including 3 university hospitals (Universiti Kebangsaan Malaysia Medical Centre (UKMMC), Hospital Universiti Sains Malaysia [HUSM] and Universiti Malaya Medical Centre [UMMC]) in Malaysia. Medical care for thalassaemia patients is available in 110 hospitals. As of the 31st of October 2019, 8964 thalassaemia patients have been registered in the MTR since year 2007.

2.1 Patient Demographics

The MTR collects a set of patients' socio-demographic details: name, identification number, date of birth, residential address, phone number, gender, ethnicity, and birth order.

2.1.1 Age Group

The mean patient age was 20.9 ± 13.3 years. The youngest patient in the registry is 3 months old, with a compound heterozygous genotype of Hb Malay and Hb Khon Kaen. The oldest patient is 88 years old, diagnosed with Haemoglobin H (HbH) disease. Patients in the 5-24.9 years old age group contributed to more than 50% of the total thalassaemia patients.

Figure 2.1: Distribution of Thalassaemia Patients in Malaysia by Age Group

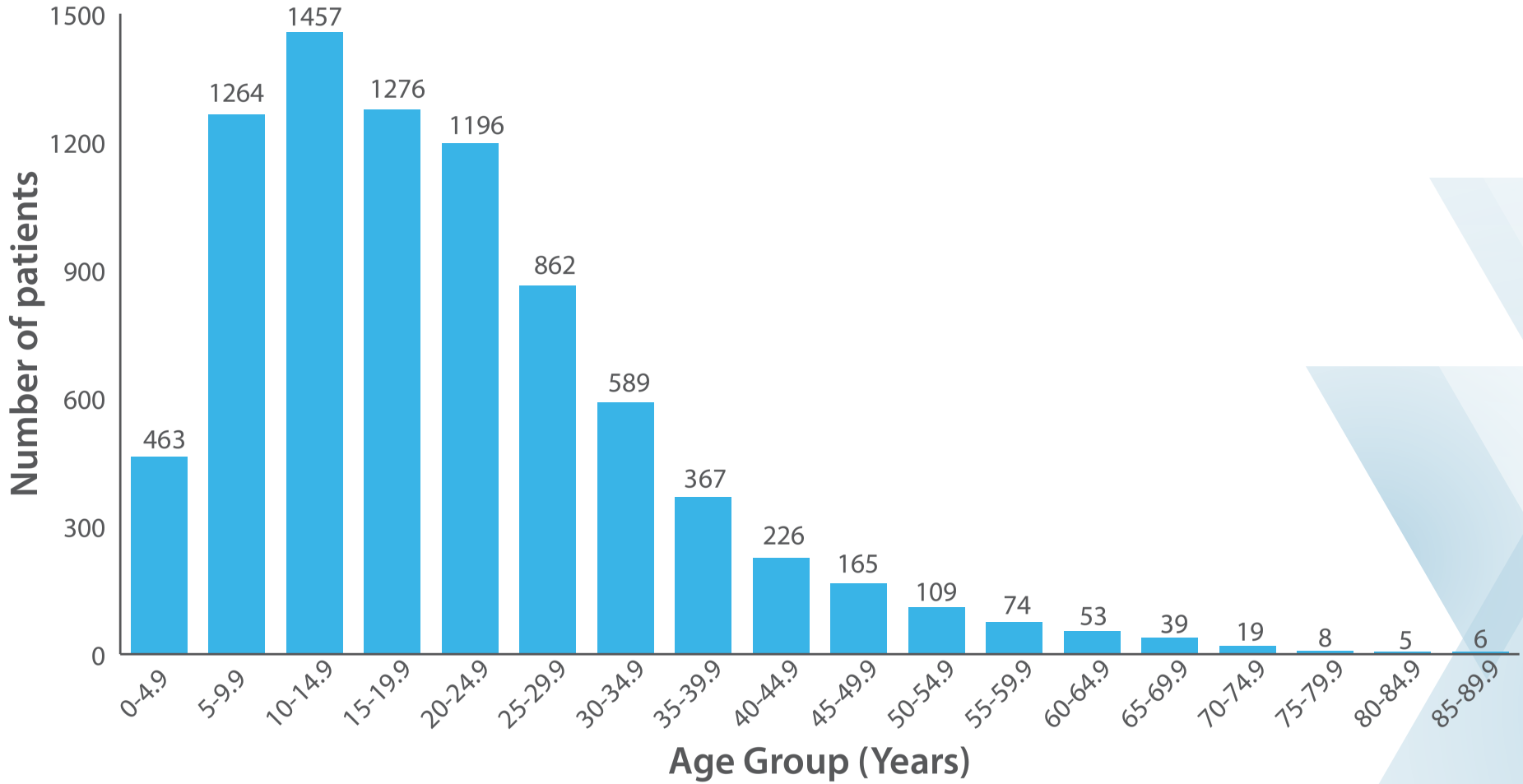
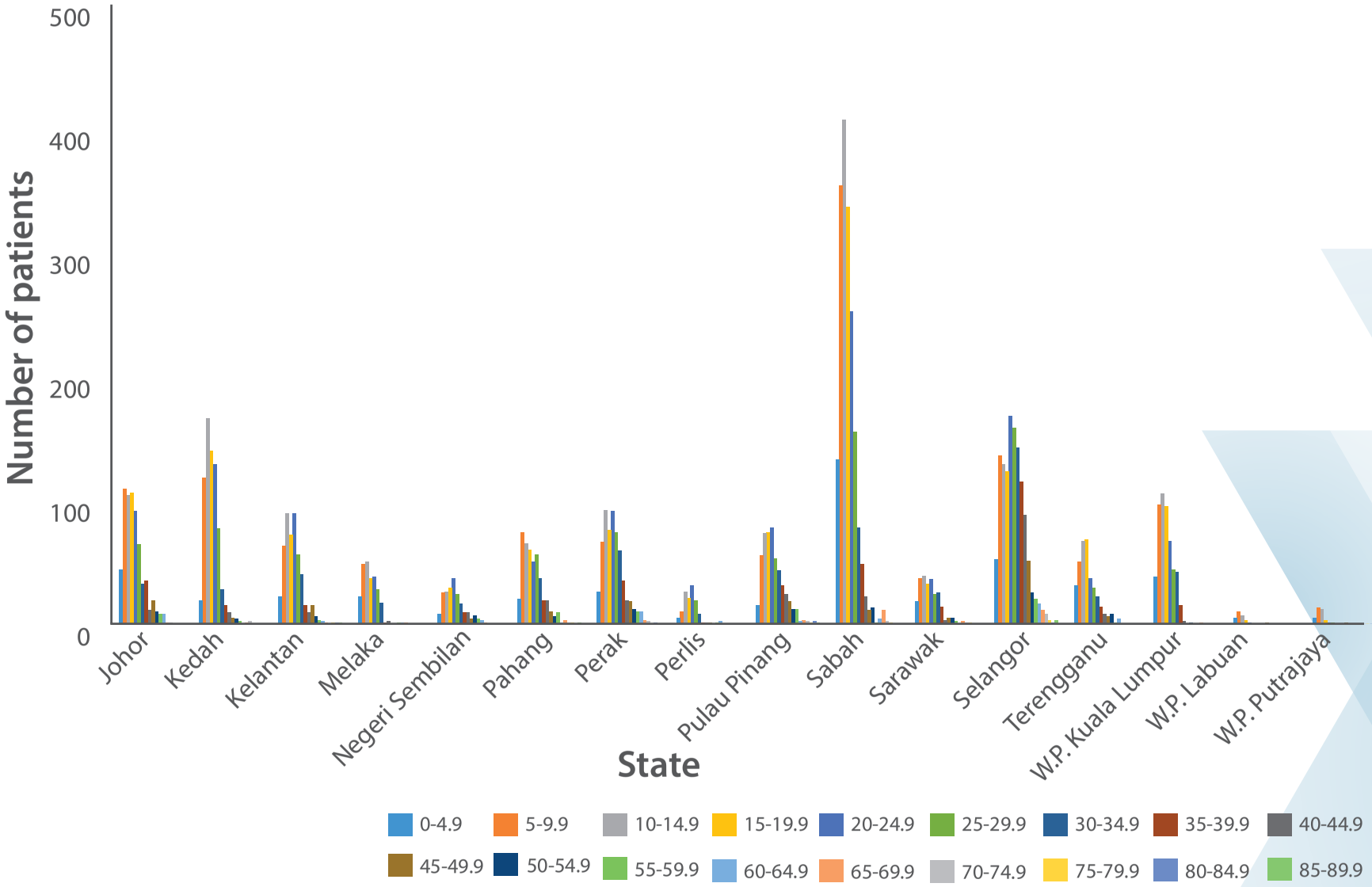


Figure 2.2: Distribution of Thalassaemia Patients in Malaysia According to Age Group by State



2.1.2 Gender

The gender distribution is almost similar between males (49.41%) and females (50.59%).

Figure 2.3: Distribution of Thalassaemia Patients in Malaysia by Gender

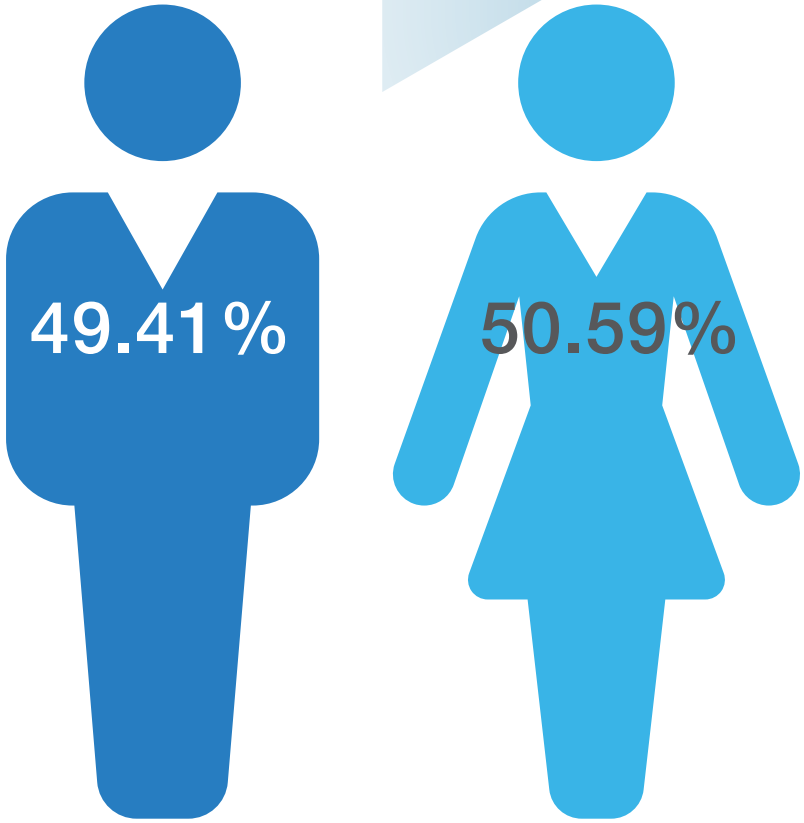
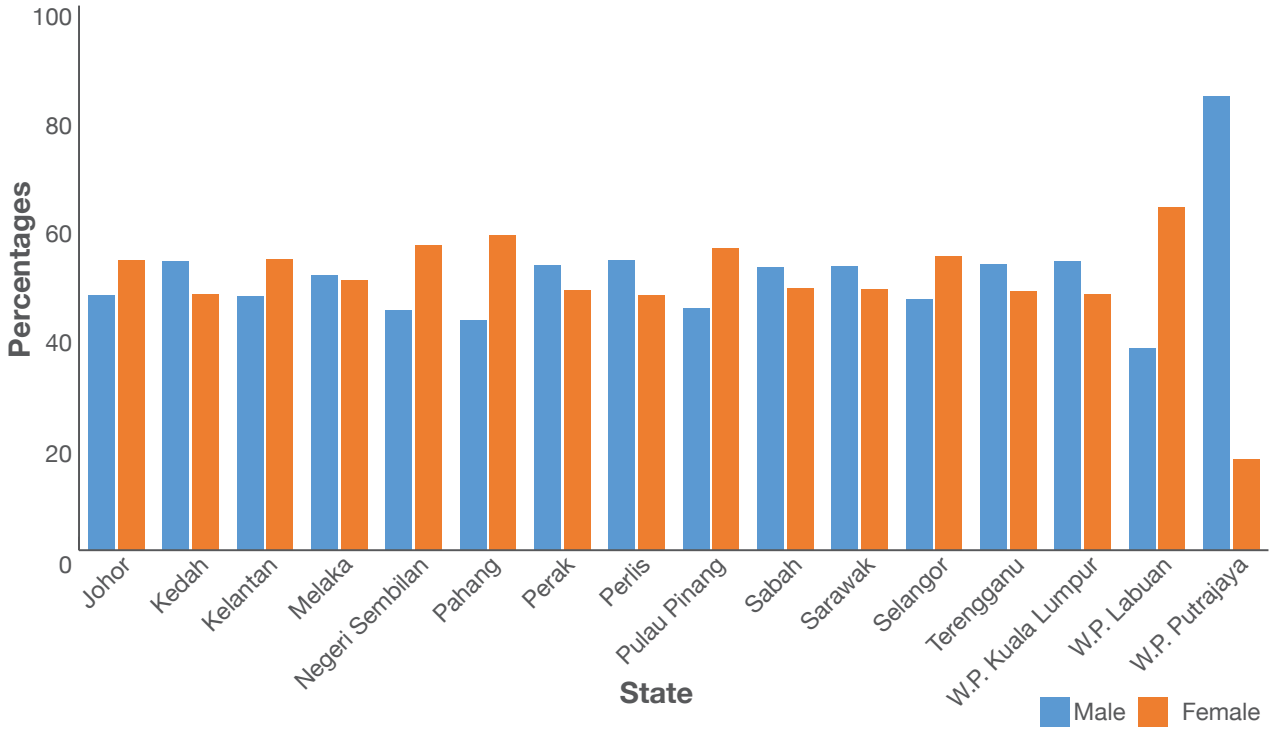


Figure 2.4: Distribution of Thalassaemia Patients in Malaysia According to Gender by State

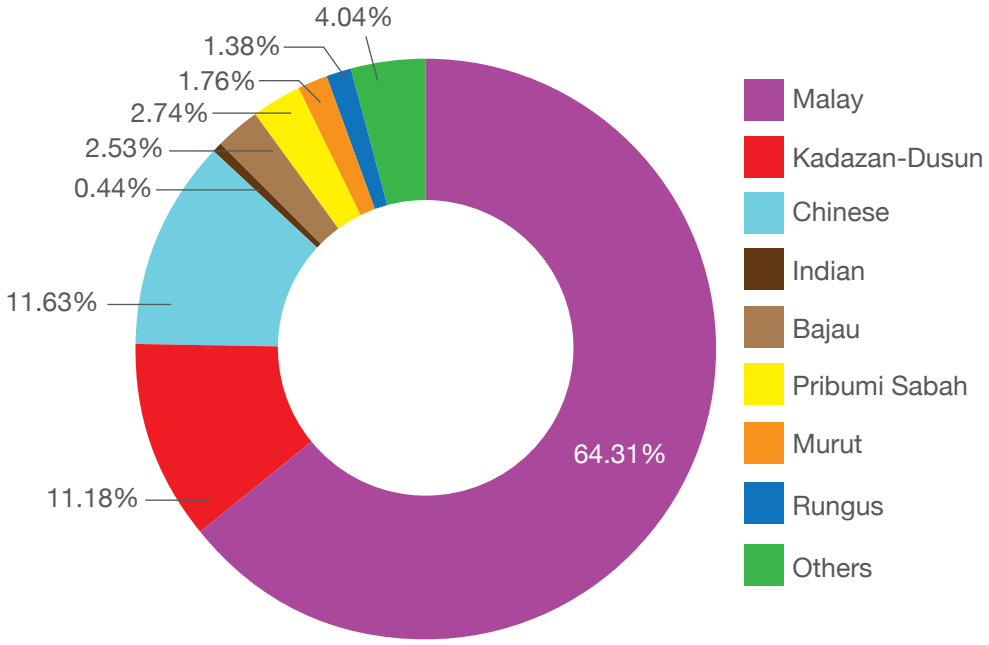


2.1.3 Ethnic Group

The three major ethnic groups in Malaysia are Malay, Chinese and Indian. However, in the states of Sabah and Sarawak, there are multiple other ethnic groups including the Kadazan-Dusun, which is the largest indigenous ethnic group in Sabah. A previous study has reported that the types and frequencies of different thalassaemia defects vary among different ethnic communities and tend to be geographically specific (Higgs et al., 1989).

Figure 2.5 shows that the Malay forms the largest group of thalassaemics with 5259 patients. This is followed by the Chinese (951) and Kadazan-Dusun (914). The 'others' status refer to other ethnic groups which are not specified. These form the remaining 330 (4.04%) patients, who are either of Orang Asli, Thai, Foreign, Iban, Pribumi Sarawak, Sino-Kadazan, Kedayan, Bidayuh, Melanau, Mixed or Kedayan ethnicities.

Figure 2.5: Distribution of Thalassaemia Patients in Malaysia by Ethnic Group



An analysis based on regional division of Peninsular Malaysia, Sabah and Sarawak showed a different pattern of patients' distribution by ethnic groups. Wilayah Persekutuan (W.P.) Kuala Lumpur and Putrajaya were grouped into Peninsular Malaysia whereby W.P. Labuan was grouped into Sabah based on geographical proximity. Sabah has the highest number of patients; predominated by the Kadazan-Dusun.

Table 2.1: Distribution of Patients by Major Ethnic Groups Based on Modified Geographical Regions

State	Total Number of Patients	Malay		Chinese		Indian		Kadazan-Dusun	
		No.	%	No.	%	No.	%	No.	%
Peninsular Malaysia	6078	5044	82.99	764	12.57	35	0.58	42	0.69
Sabah + Labuan	1856	109	5.87	96	5.17	1	0.05	868	46.77
Sarawak	244	106	43.44	90	36.89	0	0.00	4	1.64
Total	8178	5259		950		36		914	

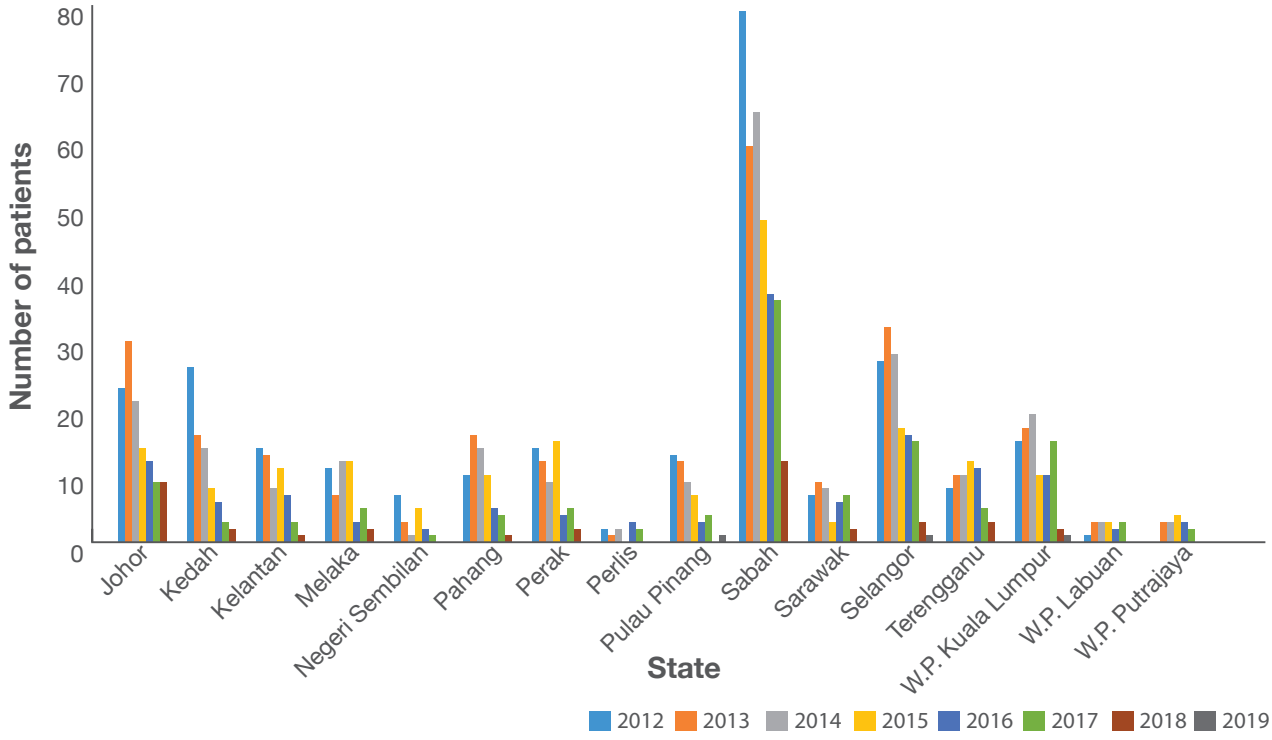
Note: Percentage (%) is calculated based on the geographic regional total.

2.2 Birth Summary

Overall, we observe a declining trend of affected births from 2015 onwards. This could likely explain the decreasing trend of total new cases between 2016 and 2018, given the clinical manifestation of TDT. From 2014 to 2018, new thalassaemia births declined steadily especially in Sabah, which could be associated with increased public awareness due to initiatives carried out by the government, in addition to screening of secondary school children.

In 2019, there were only 3 new thalassaemia births recorded, bearing in mind that diagnosis is usually only made after the first year of life following presentation of symptoms. Given the dynamic nature of the database, these patients were only registered in the MTR after diagnosis is made, instead of in the year of birth.

Figure 2.6: Number of Thalassaemia Births in Malaysia by State (2012 – 2019)



2.3 Diagnosis

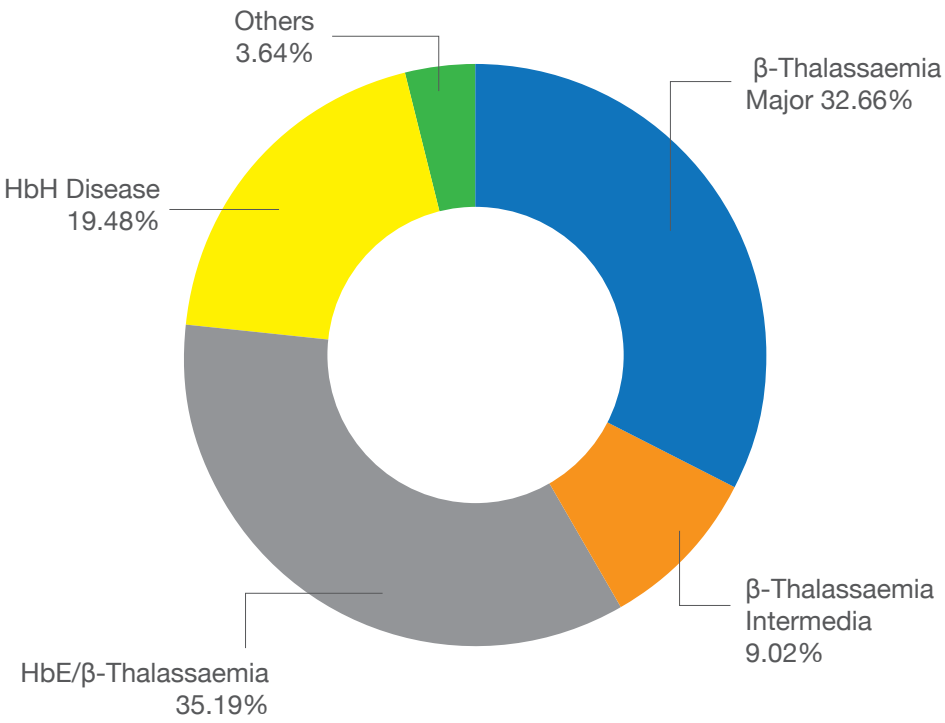
The main diagnoses of thalassaemia in the registry are β-thalassaemia major, β-thalassaemia intermedia, haemoglobin E (HbE)/β-thalassaemia and HbH disease. Other diagnoses of thalassaemia include Hb Lepore Hollandia, β-thalassaemia with Southeast Asian ovalocytosis and sickle cell β-thalassaemia.

The β-thalassaemia syndromes are much more diverse than the α-thalassaemia syndromes due to the diversity of mutations that produce defects in the β-globin gene. Patients diagnosed with thalassaemia intermedia have either a homozygous or heterozygous β-globin mutation causing a decrease in β-chain production.

Table 2.2: Distribution of Thalassaemia Patients in Malaysia by Diagnosis in 2019

Diagnosis	Number of Patients	Percentage (%)
β-Thalassaemia Major	2671	32.66
β-Thalassaemia Intermedia	738	9.02
HbE/β-Thalassaemia	2878	35.19
HbH Disease	1593	19.48
Others	298	3.64
Total	8178	100

Figure 2.7: Distribution of Patients in Malaysia by Diagnosis in 2019



The interaction of HbE with β-thalassaemia results in HbE/β-thalassaemia, an extremely heterogeneous clinical condition. HbE/β-thalassaemia is the most common form of β-thalassaemia in Southeast Asia. As depicted in Table 2.2, the HbE/β-thalassaemia forms the largest group of thalassaemia patients in Malaysia with 2878 (35.19%) patients, followed by β-thalassaemia major with 2671 (32.66%) patients, HbH disease with 1593 (19.48%) patients, β-thalassaemia intermedia with 738 (9.02%) patients, whereas the remaining 298 (3.64%) patients have other forms of thalassaemia.

Figure 2.8: Cumulative Number of Thalassaemia Patients in Malaysia According to Diagnosis by Year

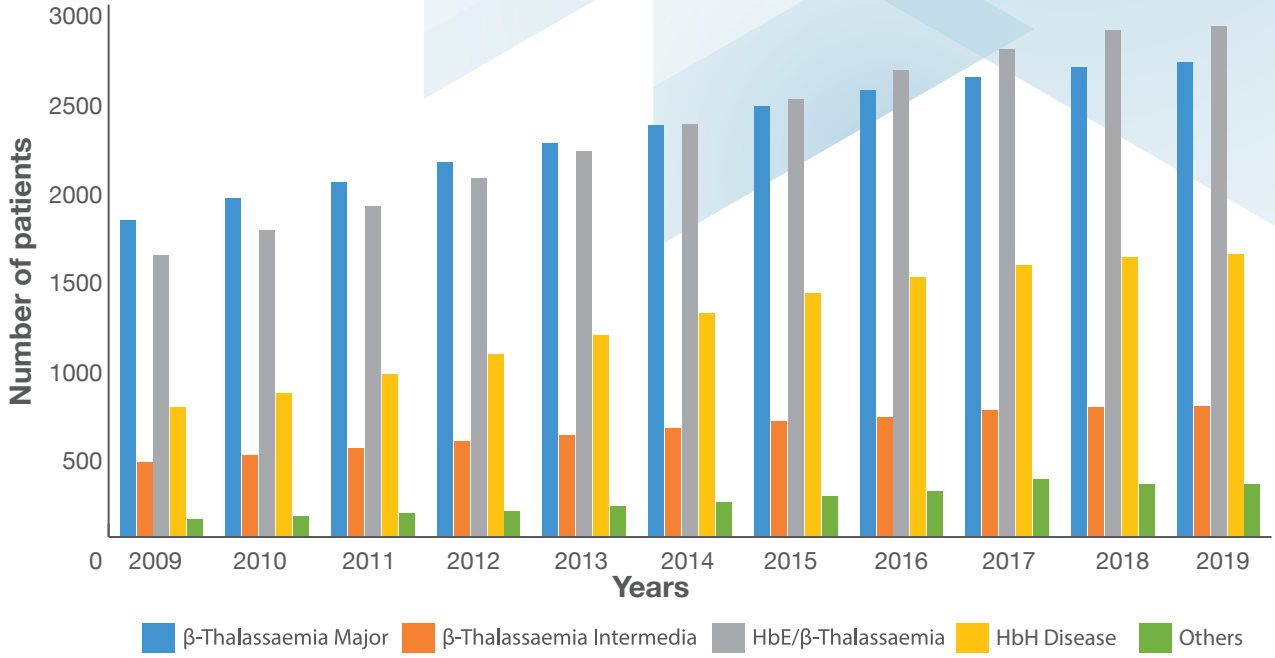


Figure 2.9: Distribution of Thalassaemia Patients in Malaysia According to Diagnosis by State

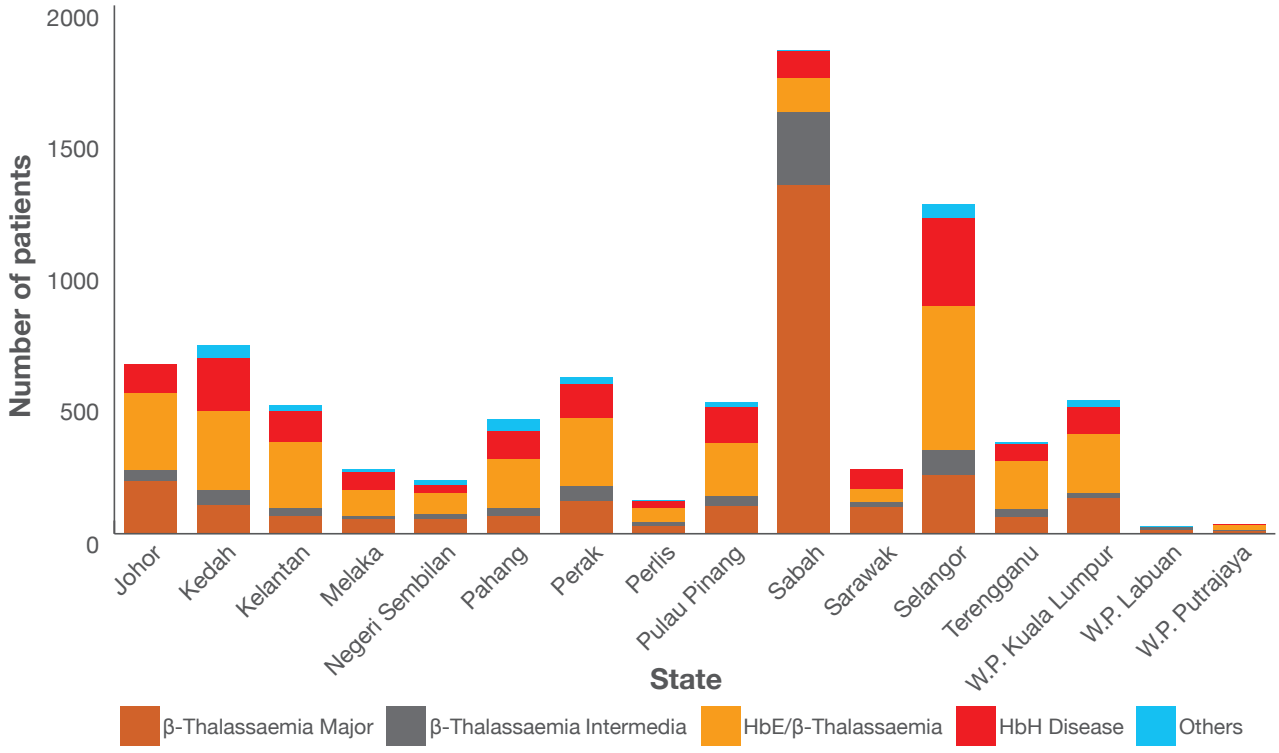
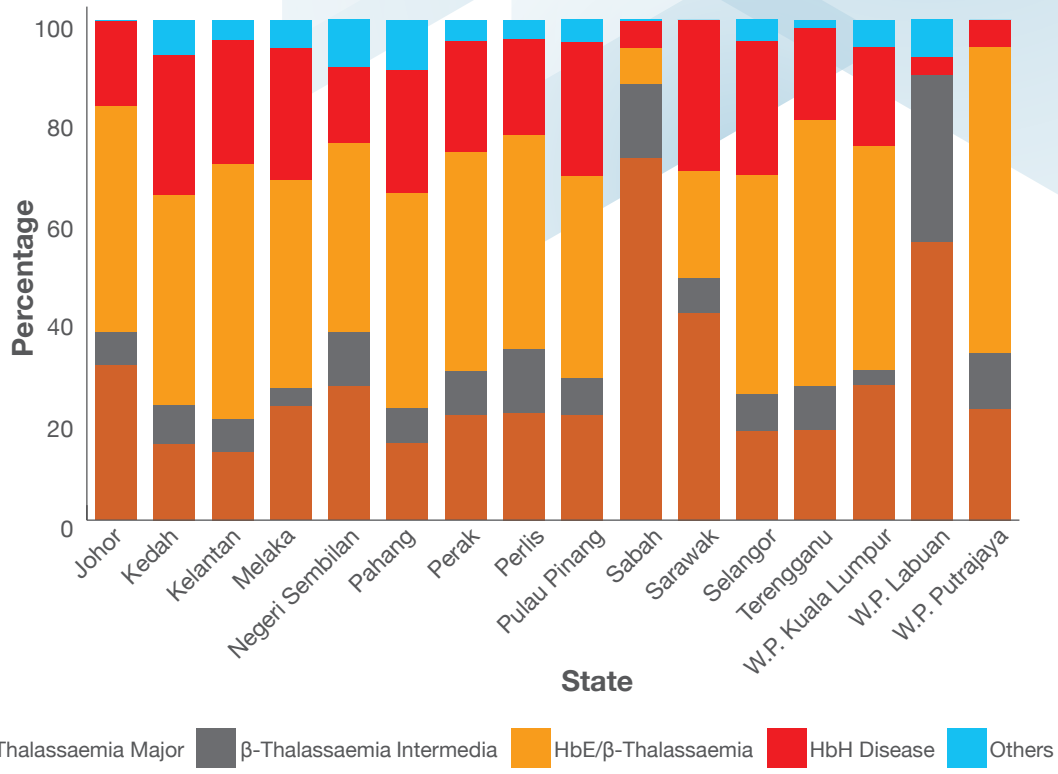


Figure 2.10: Percent Distribution of Diagnosis by State



The distribution of thalassaemia diagnoses in each state varies as shown in Figure 2.10. For actual numbers, refer to Table A4.

Figure 2.11: Distribution of Thalassaemia Patients in Malaysia According to Diagnosis by Age Group

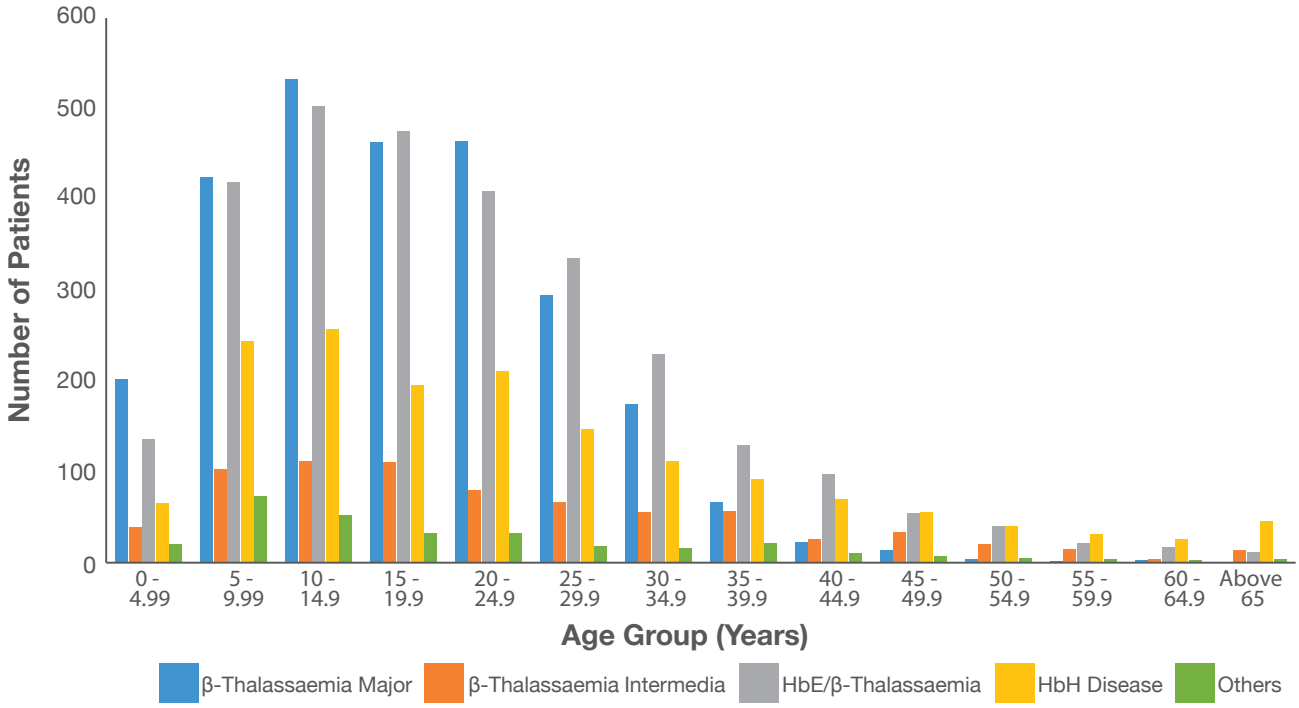


Figure 2.11 shows that most patients fall in the 10-14.9 years old age group. Out of 2671 patients with β-thalassaemia major, 2180 (81.62%) are within 5-30 years old. Likewise for HbE/β-thalassaemia and HbH disease, out of 2878 and 1593 patients, 2141 (74.39%) and 1054 (66.16%) also fall in the 5-30 years old age group, respectively.

Table 2.3: Distribution of Thalassaemia Patients in Malaysia According to Diagnosis by Ethnic Group

Diagnosis	Malay		Chinese		Indian		Kadazan-Dusun		Others	
	No.	%*	No.	%*	No.	%*	No.	%*	No.	%*
β-Thalassaemia Major	940	17.87	393	41.37	12	33.33	782	85.56	544	53.39
β-Thalassaemia Intermedia	394	7.49	72	7.58	5	13.89	90	9.85	177	17.37
HbE/ β-Thalassaemia	2550	48.49	131	13.79	7	19.44	27	2.95	163	16.00
HbH Disease	1133	21.54	325	34.21	5	13.89	11	1.20	119	11.68
Others	242	4.60	29	3.05	7	19.44	4	0.44	16	1.57
Total	5259	100.00	950	100.00	36	100.00	914	100.00	1019	100.00

*Percentages are calculated against the total for each ethnic group.

Table 2.3 demonstrates that HbE/β-thalassaemia is more common among the Malay (2550 patients, 48.49%). Meanwhile, β-thalassaemia major is more prevalent among the Chinese, Indian and Kadazan Dusun ethnicities with 393 (41.37%), 12 (33.33%), and 782 (85.56%) cases, respectively. There is a lack of information regarding the prevalence of thalassaemia in the indigenous ethnic population.

2.4 Ten Centres with the Highest Number of Thalassaemia Patients in Malaysia

Hospital Ampang in Selangor has the highest number of thalassaemia patients among the 110 participating centres. This may be attributed to the fact that Hospital Ampang is the national referral centre for adult haematology patients, besides being located in a high-density location, Klang Valley. In addition, most paediatric thalassaemia patients from Hospital Kuala Lumpur are transferred to Hospital Ampang once they transition into adulthood.

Table 2.4: Ten Centres with the Highest Numbers of Thalassaemia Patients in Malaysia

Centre	Number of Patients (n)
Hospital Ampang	731
Hospital Queen Elizabeth, Kota Kinabalu	369
Hospital Sultanah Bahiyah, Alor Setar	359
Hospital Raja Permaisuri Bainun, Ipoh	348
Hospital Sultanah Aminah, Johor Bahru	322
Hospital Wanita & Kanak-Kanak Likas, Sabah	308
Hospital Pulau Pinang	264
Hospital Tengku Ampuan Afzan, Kuantan	260
Hospital Melaka	246
Hospital Raja Perempuan Zainab II, Kota Bharu	235

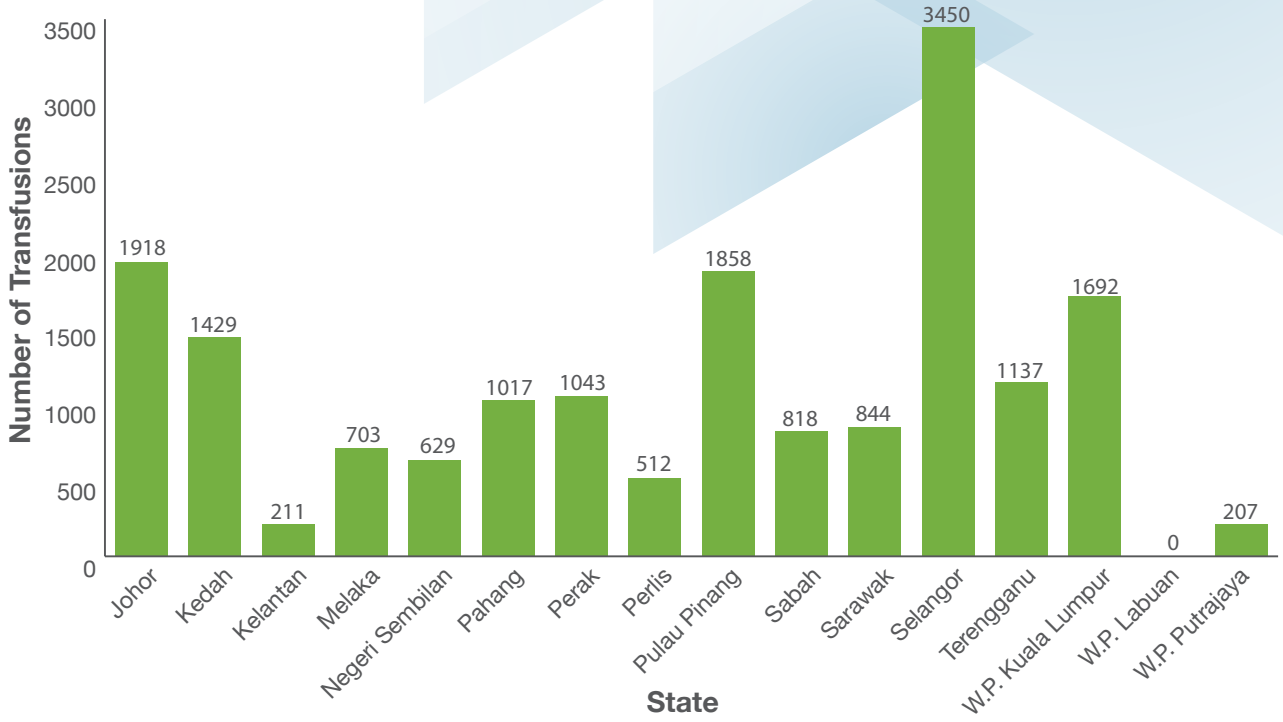
2.5 Blood Transfusion

There are 4718 TDT patients who receive regular packed red blood cell (PRBC) transfusions, every 3-4 weeks to mitigate the chronic anaemia. The remaining 3460 patients are NTDT patients. Regular blood transfusion is defined as receiving PRBC transfusion at least on a 12-weekly interval. Detailed documentation of pre-transfusion haemoglobin levels, volume of total PRBCs received in a year, transfusion reactions and alloimmunisation is necessary for high quality care.

Table 2.5: Distribution of Patients According to Transfusion Requirement by State in 2019

State	Total Number of Patients	Number of NTDT Patients	Number of TDT Patients	Percentage of TDT Patients (%)	Total Number of Transfusions
Johor	643	260	383	59.56	1918
Kedah	715	354	361	50.49	1429
Kelantan	485	229	256	52.78	211
Melaka	246	115	131	53.25	703
Negeri Sembilan	202	65	137	67.82	629
Pahang	432	215	217	50.23	1017
Perak	594	296	298	50.17	1043
Perlis	126	43	83	65.87	512
Pulau Pinang	499	226	273	54.71	1858
Sabah	1829	612	1217	66.54	818
Sarawak	244	129	115	47.13	844
Selangor	1249	595	654	52.36	3450
Terengganu	345	103	242	70.14	1137
W.P. Kuala Lumpur	506	199	307	60.67	1692
W.P. Labuan	27	8	19	70.37	0
W.P. Putrajaya	36	11	25	69.44	207
Total	8178	3460	4718	57.69	17468

Figure 2.12: Number of Transfusions by State in 2019



Note: Some missing data in a few states.

Table 2.6: Distribution of Patients According to Transfusion Requirement by Year

Year	Total Number of Patients	Number of NTDT Patients	Number of TDT Patients	Total Number of Transfusions
2019	8178	3460	4718	17468
2018	7984	3455	4529	33497
2017	7882	3392	4490	27226
2016	7605	3235	4370	21230
2015	7217	3014	4203	17570
2014	6805	2787	4018	15649
2013	6386	2554	3832	15173
2012	5973	2345	3628	15160
2011	5547	2146	3401	13011
2010	5164	1962	3202	16885

Note: Some missing data for year 2019.

2.6 Iron Chelation Therapy

Iron removal due to transfusional iron overload is accomplished with chelating drugs such as desferrioxamine (DFO), deferriprone (DFP) and deferasirox (DFX) or a combination of these chelating agents. Effective chelation therapy in chronically transfused patients is achieved when iron chelators remove sufficient amount of iron, proportionate to that accumulated in the body from blood transfusions, and maintain the body iron load at a non-toxic level (Chonat and Quinn, 2018; Mobarra et al., 2016). Recent clinical studies applying MRI technology reported that all 3 iron chelators have established efficacy in significantly reducing iron loads from the liver and heart. Nevertheless, the indications for iron chelation therapy in patients should be individualised and modified at patient follow-ups, as different iron chelators may be suitable for different iron overload profiles (Taher and Cappellini, 2018).

DFO is administered at a regular dose of 20–50 mg/kg/day, subcutaneously using a portable infusion pump. Higher dosage up to 60 mg/kg/day is occasionally prescribed for patients with higher body iron stores. DFP is the first oral iron chelator to be used for transfusional iron overload in patients with TDT, when DFO therapy is contraindicated or inadequate. DFP is given three times daily due to its short half-life. DFX, a recent oral iron chelator, is taken once-daily at dose of 20–40 mg/kg/day. DFX has a longer plasma half-life of 16–18 hours, and is predominantly excreted in biliary secretions.

A total of 5084 out of 8178 (62.17%) Malaysian thalassaemia patients are on iron chelation therapy. Table 2.7 shows that 74.61% of patients are on iron chelation monotherapy while the rest are on combination therapy. DFX is usually prescribed to chelator-naïve young patients and for those who could not tolerate other chelating agents. The most common combination therapy prescribed was DFO + DFP in 955 (18.78%) patients.

Table 2.7: Distribution of Thalassaemia Patients in Malaysia Based on Iron Chelator(s) Used

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	876	17.23
DFP only	1268	24.94
DFX only	1649	32.44
DFO + DFP	955	18.78
DFP + DFX	110	2.16
DFO + DFX	187	3.68
DFO + DFP + DFX	39	0.77
Total	5084	100.00

Figure 2.13: Breakdown of Patients According to Iron Chelator(s) Used by State

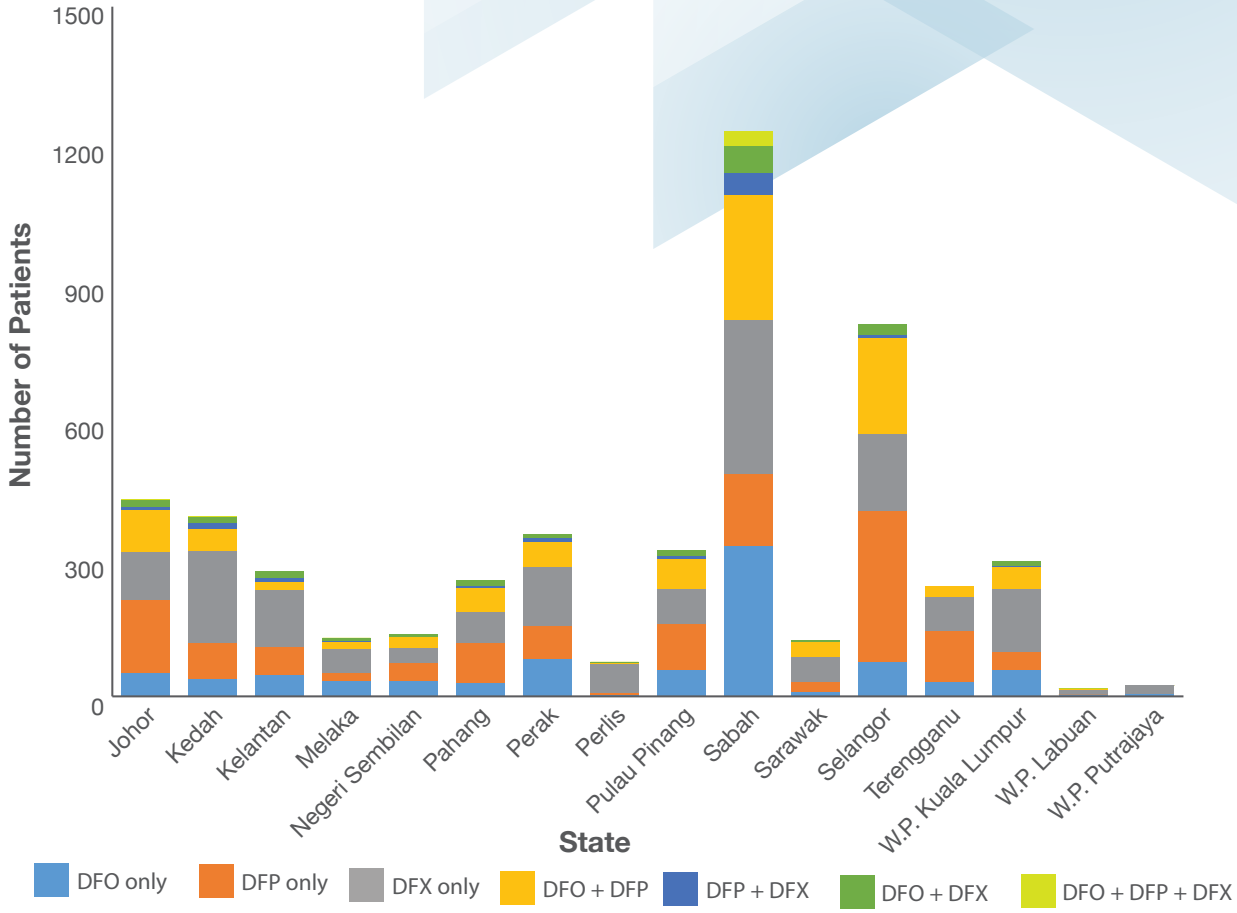
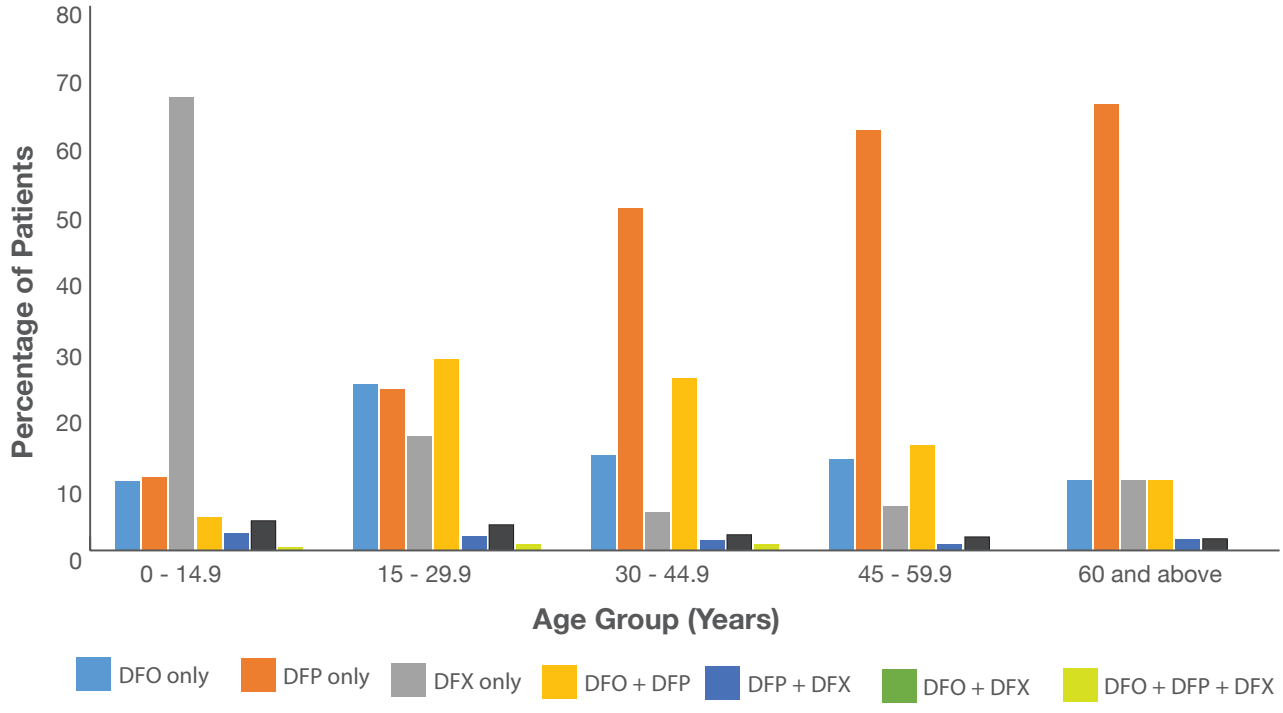


Figure 2.14: Breakdown of Patients According to Iron Chelator(s) Used by Age Group



2.7 Serum Ferritin Level Monitoring

Chronic iron overload is the most challenging complication associated with chronic blood transfusions. TDT patients will progressively develop clinical manifestation unless treated promptly and adequately. Therefore, iron status should be accurately and continuously assessed to evaluate its severity, the need for treatment initiation and for monitoring the chelation therapy. Although serum ferritin levels provide a convenient method to approximate iron overload in thalassaemics, it is not a reliable indicator of total body iron, liver overload or cardiac overload, since serum ferritin is an acute phase reactant and its levels may change for a variety of non-iron related reasons. However, the trend in serum ferritin levels could be a useful surrogate marker of total body iron level in patients. The more sensitive and specific parameter to measure tissue iron overload is liver and cardiac MRI T2* (Chirico et al., 2014).

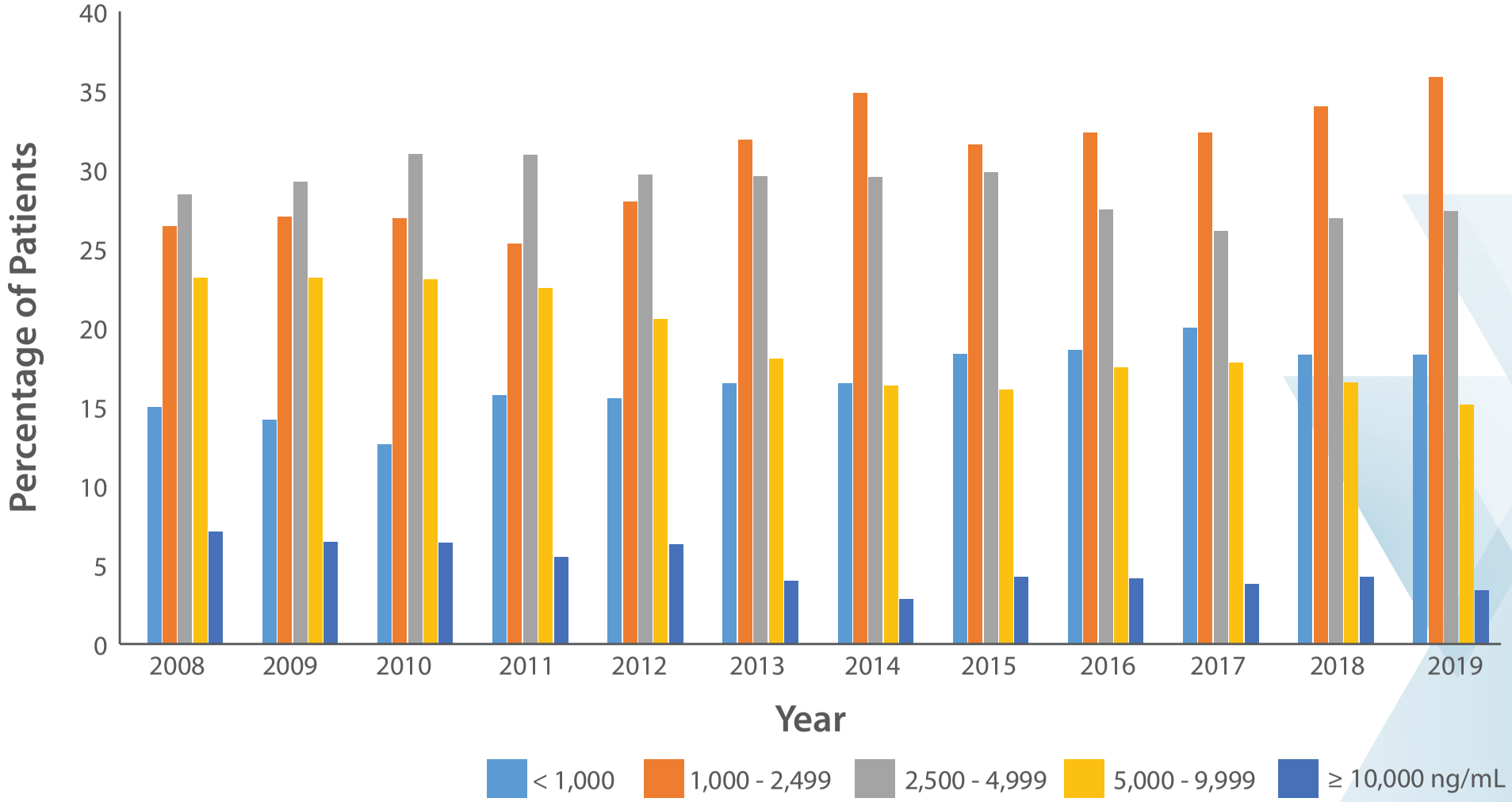
We observe a decreasing number of patients with serum ferritin levels beyond 5,000 ng/mL and more patients now have serum ferritin levels of below 2500 ng/mL. Although the trend of serum ferritin levels in TDT patients are not captured by the registry, this observation most likely indicates an improvement in iron overload management and could translate to better long-term outcomes and preservation of vital organ functions.

Table 2.8: Latest Serum Ferritin Levels (ng/mL) Based on Available Data in 2019

State	Total	< 1000		1000-2499		2500-4999		5000-9999		≥10,000	
		No.	%*	No.	%*	No.	%*	No.	%*	No.	%*
Johor	315	49	15.56	114	36.19	97	30.79	47	14.92	8	2.54
Kedah	240	38	15.83	97	40.42	64	26.67	30	12.50	11	4.58
Kelantan	91	10	10.99	40	43.96	34	37.36	6	6.59	1	1.10
Melaka	71	11	15.49	25	35.21	16	22.54	15	21.13	4	5.63
Negeri Sembilan	103	15	14.56	33	32.04	31	30.10	20	19.42	4	3.88
Pahang	149	41	27.52	63	42.28	30	20.13	14	9.40	1	0.67
Perak	243	50	20.58	83	34.16	75	30.86	27	11.11	8	3.29
Perlis	53	10	18.87	22	41.51	12	22.64	8	15.09	1	1.89
Pulau Pinang	226	43	19.03	87	38.50	58	25.66	33	14.60	5	2.21
Sabah	302	48	15.89	67	22.19	77	25.50	81	26.82	29	9.60
Sarawak	103	17	16.50	47	45.63	23	22.33	14	13.59	2	1.94
Selangor	511	83	16.24	169	33.07	155	30.33	85	16.63	19	3.72
Terengganu	222	67	30.18	77	34.68	49	22.07	26	11.71	3	1.35
W.P. Kuala Lumpur	247	45	18.22	105	42.51	66	26.72	29	11.74	2	0.81
W.P. Labuan	15	2	13.33	2	13.33	6	40.00	4	26.67	1	6.67
W.P. Putrajaya	21	3	14.29	13	61.90	4	19.05	1	4.76	0	0.00
Total	2912	532	18.27	1044	35.85	797	27.37	440	15.11	99	3.40

*Percentage(%) is calculated according to the number of patients in each respective state.

Figure 2.15: Distribution of Thalassaemia Patients in Malaysia According to Serum Ferritin Levels (ng/mL) by Year



2.8 Splenectomy

The most common reason for performing splenectomy is hypersplenism, which increases blood transfusion requirements and prevents adequate control of body iron level with chelation therapy. Splenectomy is usually indicated in patients whose yearly blood transfusion requirements exceed 200-250 mL packed cells/kg body weight with hypersplenism (with or without the presence of splenomegaly-related complications such as cytopenia, pain or risk of rupture). Hypersplenism may be avoided by adequate blood volume and regular blood transfusions. Recently, it has been reported that many patients reaching adolescence did not require splenectomy. Splenectomy should be delayed until the age of 5 years or later to avoid the risk of post-splenectomy infection (Chonat and Quinn, 2017). Out of the total 8964 patients, 1282 (14.30%) had undergone splenectomy. The majority of splenectomy was performed in patients diagnosed with HbE/β-thalassaemia and β-thalassaemia major.

Table 2.9: Distribution of Splenectomised Patients in Malaysia by State

State	Total Number of Patients	Number of Patients who Underwent Splenectomy	Percentage (%)
Johor	679	107	15.76
Kedah	747	119	15.93
Kelantan	519	110	21.19
Melaka	251	30	11.95
Negeri Sembilan	212	26	12.26
Pahang	480	115	23.96
Perak	641	78	12.17
Perlis	140	20	14.29
Pulau Pinang	514	64	12.45
Sabah	2148	220	10.24
Sarawak	265	32	12.08
Selangor	1331	231	17.36
Terengganu	373	64	17.16
W.P. Kuala Lumpur	596	63	10.57
W.P. Labuan	31	1	3.23
W.P. Putrajaya	37	2	5.41
Total	8964	1282	

*Percentage(%) is calculated according to the number of patients in each respective state.

2.9 Patients' Vital Status

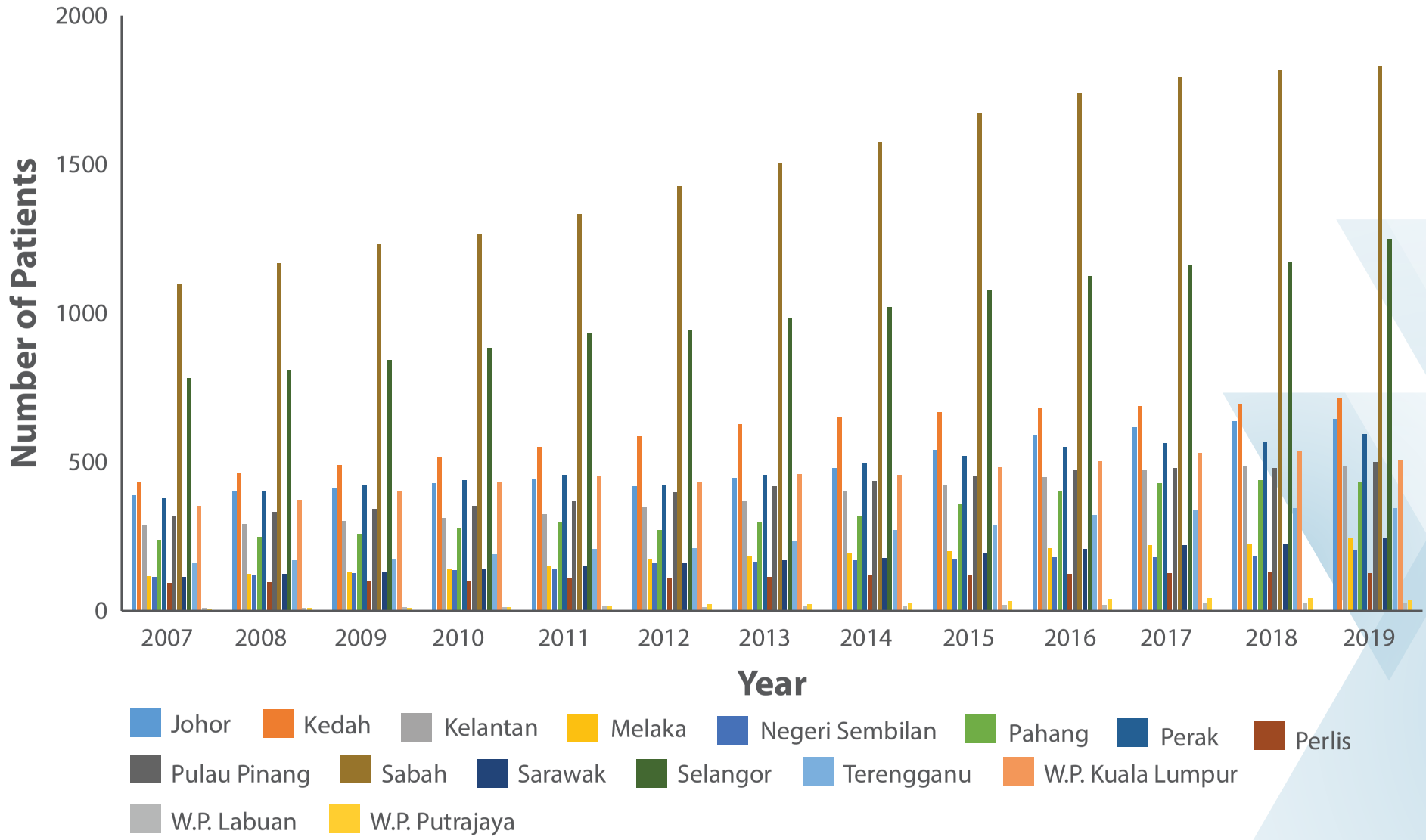
In the Registry, patients are categorised as alive and on active treatment, cured by stem cell transplant, lost to follow-up, deceased or transferred to another centre. Those who are alive and on active treatment and those cured by stem cell transplant were added together as total number of living patients, as shown in Table 2.10.

Table 2.10: Distribution of Patients by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	7201
Cured by Stem Cell Therapy	147
Total Living Patients	7348
Lost to Follow-up	830
Total	8178
Cumulative Reported Deaths	786
Total	8964

Similar to 2018, Sabah has the highest cumulative number of registered thalassaemia patients in Malaysia, at 1829 (22.36%) patients. This is followed by Selangor, Kedah and Johor at 1249, 715 and 643 patients, respectively. Meanwhile, Wilayah Persekutuan Labuan reported the lowest number of cases at 27 patients.

Figure 2.16: Distribution of Thalassaemia Patients in Malaysia According to State by Year (2007-2019)



2.10 Mortality Data

Morbidity and mortality in thalassaemia patients are mainly due to complications from chronic iron overload, which have led to several fatal events. The total number of deaths recorded in the MTR is 786 (8.77%) out of 8964 patients since 2007. The data demonstrated that cardiac-related complications remain the leading cause of death. The cumulative reported number of deaths by state is presented in Table 2.11. Sabah has the highest number of deaths followed by Kuala Lumpur, Selangor and Perak, respectively. Meanwhile, Negeri Sembilan, Melaka, Putrajaya and Labuan recorded fewer number of deaths.

Out of the 786 deaths recorded, 75 had incomplete data. The leading causes of death in 711 deceased patients with verifiable data in Malaysia were cardiac-related causes (292 cases, 41.07%) and infection (278 cases, 39.10%).

Table 2.11: Cumulative Reported Deaths by State from 2007 until October 2019

State	Number of Patients
Johor	36
Kedah	32
Kelantan	34
Melaka	5
Negeri Sembilan	10
Pahang	48
Perak	47
Perlis	14
Pulau Pinang	15
Sabah	318
Sarawak	21
Selangor	82
Terengganu	28
W.P. Kuala Lumpur	90
W.P. Labuan	4
W.P. Putrajaya	1
Total	786

Our findings are similar to the reported studies in other countries, in which the major cause of death was cardiac complications or dysfunctions. Previous studies reported that heart failure in these patients could be due to constrictive pericarditis or cardiomyopathy secondary to iron overload (Borgna-Pinatti et al., 2004; Ladis et al., 2005; Fucharoen et al., 2011).

Other causes of death include motor vehicle accidents (2.81%), liver disease (2.67%) and malignancy (2.67%). Another category of death recorded in this registry is died at home/brought in dead without post mortem.

Table 2.12: Cumulative Causes of Death Since 2007

Causes of Death	Number of Patients	Percentage (%)
Cardiac-Related Causes	292	41.07
Infection	278	39.10
Motor Vehicle Accidents	20	2.81
Liver Disease	19	2.67
Malignancy	19	2.67
Central Nervous System Event	12	1.69
Endocrine Complications	13	1.83
Renal Complications	8	1.13
Thalassaemia	6	0.84
Bone Marrow Transplant Complications	5	0.70
Surgical Complications	5	0.70
Thrombosis	3	0.42
Died at Home/Brought in Dead to Hospital	17	2.39
Others	14	1.97
Total	711	100.00

Figure 2.17: Number of Reported Deaths by Year (2009 – 2019)

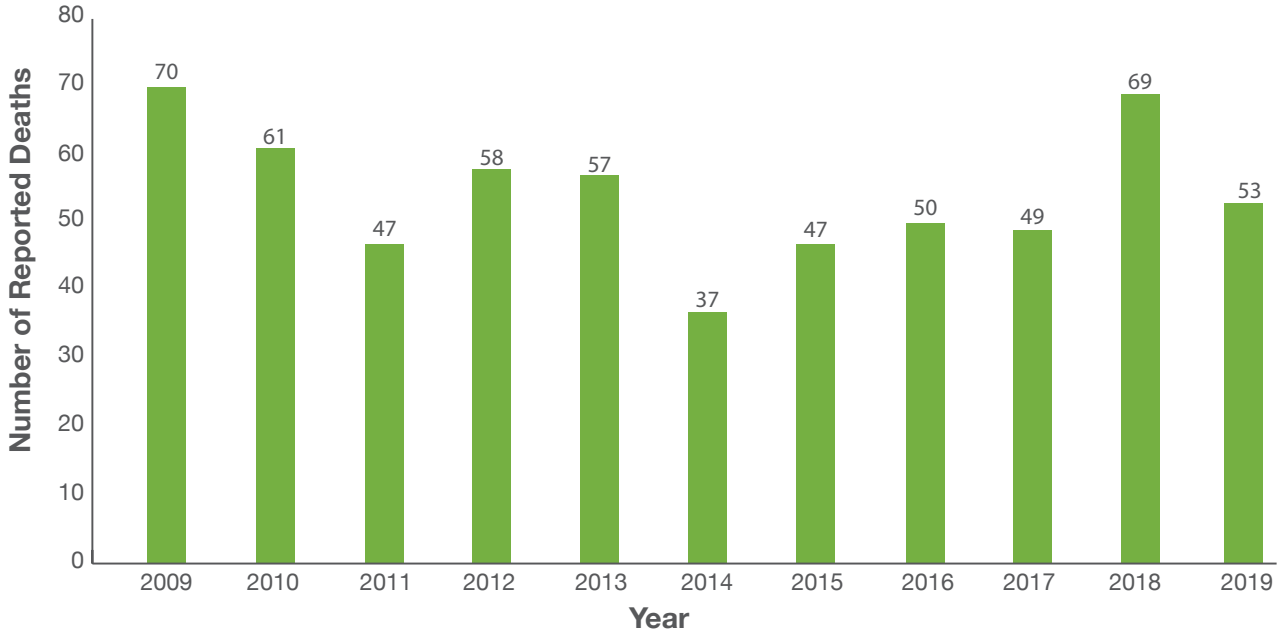
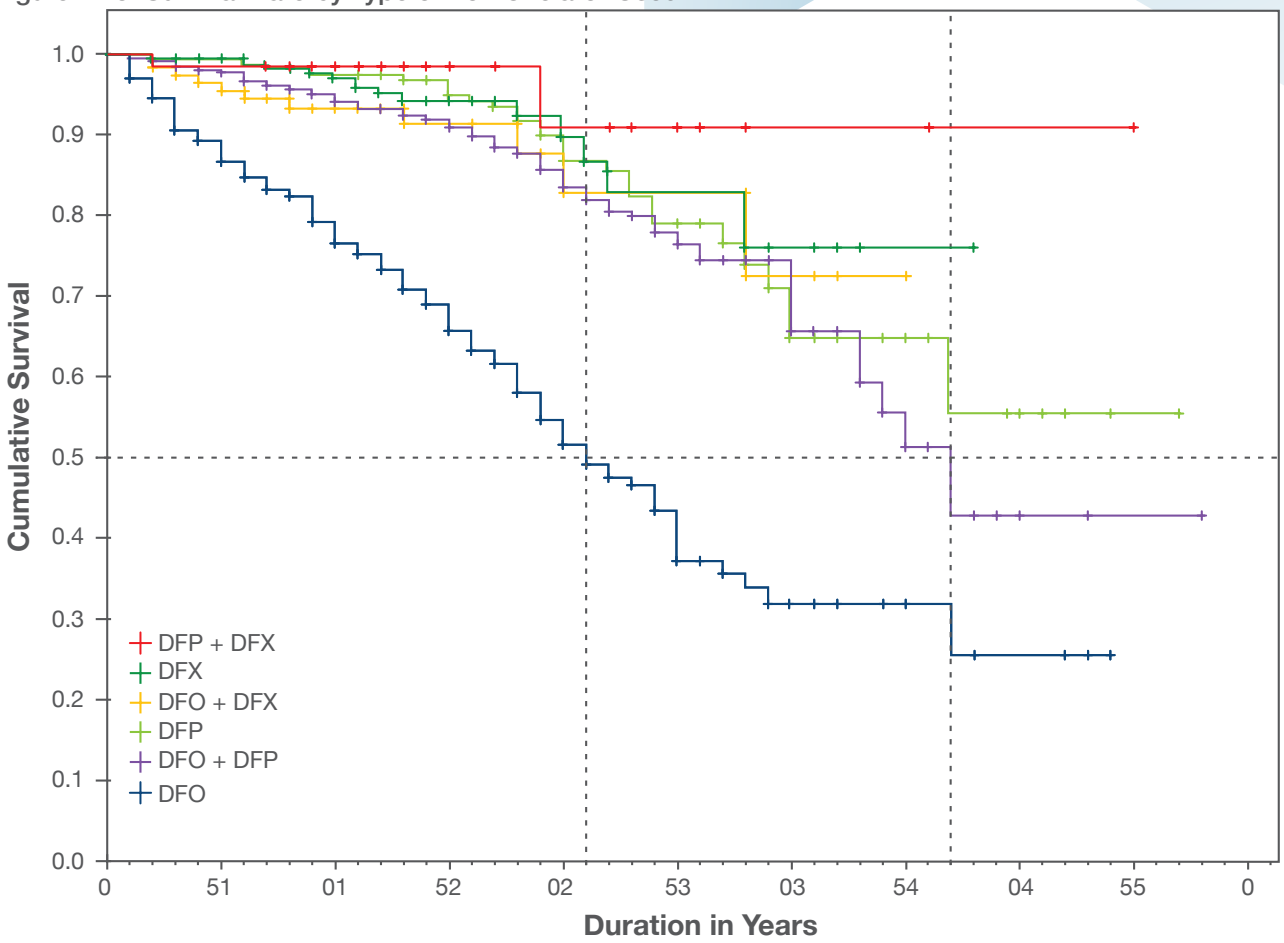


Figure 2.17 demonstrates the annual thalassaemia deaths over the past 10 years.

2.11 Treatment Analysis

2.11.1 Overall survival of β-thalassaemia major patients by type of chelators

Figure 2.18: Survival Rate by Type of Iron Chelator Used



The mean survival duration is 37.48 (standard deviation, SD ±1.60) years for β-thalassaemia major patients on DFP, 33.70 (SD ±1.20) years for patients on DFX, and 30.44 (SD ±1.35) years for DFO + DFX from diagnosis. The median survival duration for TDT patients on DFO monotherapy and DFO + DFP are 21 years and 37 years from diagnosis, respectively. The findings of the overall survival must be interpreted cautiously, as only the currently used iron chelator(s) are being analysed. The type of chelator prescribed earlier is not captured by the registry. Analysis on the effect of gender, city of residence and other types of thalassaemia were not performed, and we hope this could be done in the subsequent report.

3 National Conclusions and Observations

A few observations can be made from this report:

1. The total cumulative numbers of thalassaemia patients has increased to 8178 compared to 7984 in year 2018. This is contributed by improved overall survival and more patients being registered into the MTR.

2. The largest number of registered patients came from the state of Sabah, followed by Selangor, Kedah, Johor and Perak.

3. Most patients in Sabah have β-thalassaemia major, whereas patients in Peninsular Malaysia predominantly have HbE/β-thalassaemia (either TDT or NTDT). These findings can facilitate healthcare planning and resource distribution.

4. The most common genotype abnormality found among β-thalassaemia major patients in Sabah is the Filipino (FIL) ~45-kb deletion. In Peninsular Malaysia, HbE/β-thalassaemia is the most common diagnosis found among Malay patients. Early identification of the genotypic mutations allows better prediction of phenotypic manifestation and severity of the disease. A complete molecular diagnosis to identify the primary/secondary alleles of thalassaemia and also to identify gene modifiers are important for an early and accurate diagnosis (George, 2013).

5. Number of new births and newly diagnosed cases from 2015 until present have decreased (Hishamshah et al., 2020). From 2014 to 2018, new thalassaemia births have declined steadily especially in Sabah. This is may be associated with increased public awareness due to initiatives carried out by the government, in addition to screening of secondary school children.

6. The number of patients with serum ferritin levels beyond 5,000 ng/mL is decreasing, and more patients now have serum ferritin levels below 2500 ng/mL. This observation most likely indicate an improvement in iron overload management and monitoring.

7. The most common iron chelator used is oral DFX monotherapy (32.44%). DFX is especially prescribed in chelator-naïve patients.

8. Newly added variables on end organ complications in the MTR are still in progress; therefore, a relevant analysis will be carried out in the future.

9. The cumulative number of patients cured by stem cell transplant is 147, and expected to increase as the transplant services are expanded in the country.

10. The accumulated number of deaths since 2007 is 786. The two most common causes of death are cardiac complications (292 cases) and infection (278 cases).

4 Johor

4.1 Introduction

Johor is located at the southernmost region of Peninsular Malaysia. The total population of Johor is about 3.764 million people (Department of Statistics Malaysia, 2019).

There are 12 hospitals in the state of Johor. Six are hospitals with specialists, namely Hospital Sultanah Aminah, Johor Bahru (HSAJB), Hospital Sultan Ismail (HSI), Hospital Enche' Besar Hajjah Khalsom, Kluang (HEBHK), Hospital Sultanah Nora Ismail, Batu Pahat (HSNI), Hospital Pakar Sultanah Fatimah, Muar (HPSF) and Hospital Segamat. The other five hospitals, namely Hospital Temenggong Seri Maharaja Tun Ibrahim, Kulai (HTSMTI), Hospital Kota Tinggi, Hospital Mersing, Hospital Pontian and Hospital Tangkak are district hospitals, but only Hospital Mersing handles thalassaemia patients. Another hospital, Hospital Permai, solely treats psychiatric patients.

Hospital Sultanah Aminah is a haematology centre and caters for both paediatrics and adults with thalassaemia, whereas Hospital Sultan Ismail is a paediatric haematology-oncology centre treating all haematological problems, including thalassaemia in patients below 18 years old.

All blood products received by the thalassaemia patients had undergone nucleic acid testing (NAT) at Pusat Darah Negara in order to reduce the risk of transfusion-transmitted infection (TTI). All adult thalassaemia patients in Hospital Sultanah Aminah received pre-storage filtered blood. Meanwhile, paediatric patients only received bed-side filtered blood (if filters available).

4.2 Patient Demographics

Majority of patients in Johor received treatment at Hospital Sultanah Aminah (50.08%) and Hospital Sultan Ismail (14.46%), as both centres are located in Johor Bahru (which has the highest population in the state at about 1.3 million). These centres are also the referral centres for thalassaemia cases, as shown in Table 4.1.

Table 4.1: Distribution of Patients by Centre in Johor

Centre	Number of Patients	Percentage %
Hospital Sultanah Aminah	322	50.08
Hospital Sultan Ismail	93	14.46
Hospital Pakar Sultanah Fatimah	89	13.84
Hospital Enche' Besar Hajjah Kalsom	43	6.69
Hospital Segamat	36	5.60
Hospital Mersing	30	4.67
Hospital Sultanah Nora Ismail	30	4.67
Hospital Tangkak	0	0.00
Hospital Temenggung Seri Maharaja Tun Ibrahim	0	0.00
Hospital Pontian	0	0.00
Hospital Kota Tinggi	0	0.00
Total	643	100.00

As shown in Table 4.2, there are 562 living patients in 2019 and 14 were cured by stem cell transplantation and another 67 patients were lost to follow-up. Meanwhile, a total of 36 deceased patients with different causes of death has been reported since 2009 in Johor (Table 4.3). There were 3 thalassaemia patients deaths reported in 2019, due to central nervous system, tumour and cardiac-related causes (Table 4.4).

Table 4.2: Distribution of Patients by Vital Status in Johor

Vital Status	Number of Patients
Alive	562
Cured by Stem Cell Therapy	14
Total	576
Lost to Follow-up	67
Total	643
Deaths in 2019	3
Cumulative Reported Deaths (from 2009 to 2019)	36

Table 4.3: Cumulative Causes of Death in Johor Since 2007

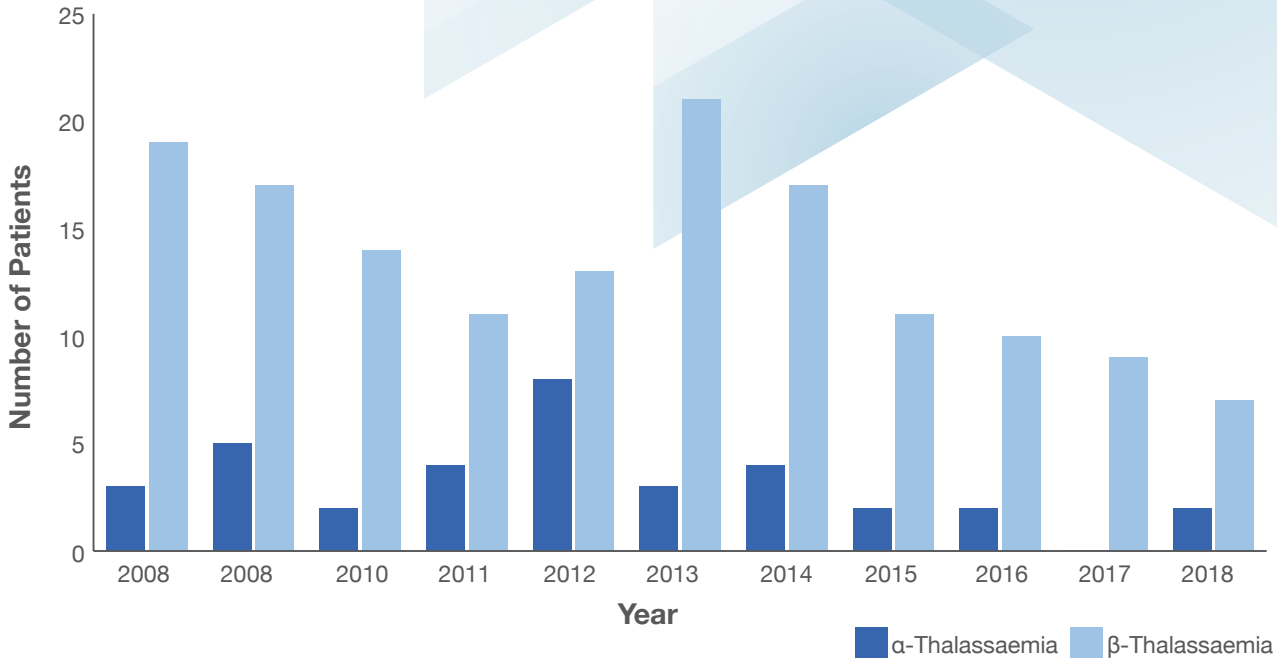
Causes of Death	Number of Patients	Percentage (%)
Infections	19	52.78
Cardiac Causes	8	22.22
Thalassaemia	1	2.78
Malignancy	3	8.33
Motor Vehicle Accident	1	2.78
Central Nervous System Event	2	5.56
Surgical Complications	1	2.78
Others	1	2.78
Total	36	100.00

Table 4.4: Thalassaemic Deaths in Johor in 2019

Age	Gender	Centre	Diagnosis	Cause of Death	Date of Death
12	Male	Hospital Sultanah Aminah	β-thalassaemia intermedia	Severe intracranial bleeding (ICB)	30th April 2019
17	Male	Hospital Sultanah Aminah	HbE/ β-thalassaemia	Brain tumour	30th July 2019
68	Female	Hospital Enche' Besar Hajjah Kalsom	HbE/ β-thalassaemia	Advanced heart failure	30th Jan 2019

A total of 12 new cases were reported in Johor in 2019. Nine cases were new births reported in 2018 (7 patients from HSAJB, and 2 patients from HSI). Most of the new births are β-thalassaemia major cases. Figure 4.1 illustrates the trend of new thalassaemia births in Johor. Overall, no significant changes in the new births trend were observed in over 10 years.

Figure 4.1: Distribution of New Thalassaemia Births in Johor According To Year And Diagnosis



4.2.1 Age Group

Thalassaemia patients aged 5 to 20 years constituted the highest number of patients in Johor (Figure 4.2). In all age groups except those above 60 years old, most patients were diagnosed with HbE/β-thalassaemia (Table 4.5). The youngest patient in Johor is an 11-month-old boy diagnosed with β-thalassaemia major from HSI, and the eldest patient is a 70 year-old male with HbH disease under follow-up in HSA.

Figure 4.2: Distribution of Patients in Johor by Age Group

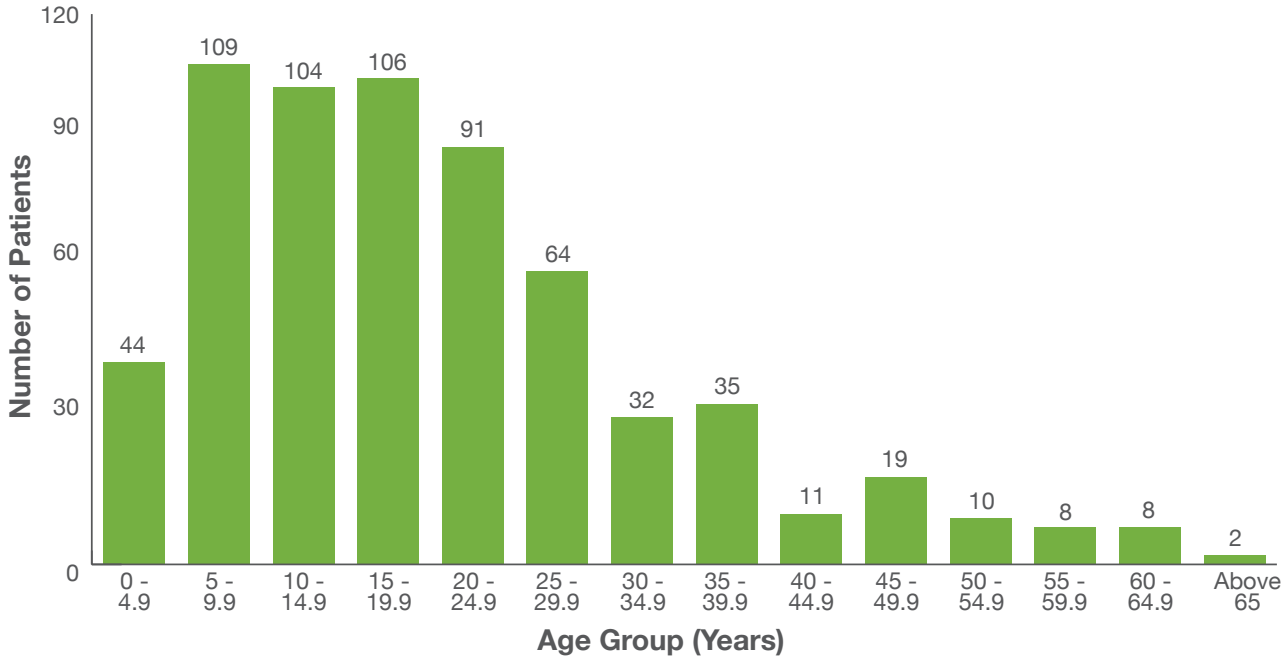


Table 4.5: Distribution of Patients in Johor According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	257	β-Thalassaemia Major	72	28.02
		β-Thalassaemia Intermedia	11	4.28
		HbE/β-Thalassaemia	119	46.30
		HbH Disease	53	20.62
		Others	2	0.78
15 - 29.9	261	β-Thalassaemia Major	98	37.55
		β-Thalassaemia Intermedia	9	3.45
		HbE/β-Thalassaemia	122	46.74
		HbH Disease	32	12.26
		Others	0	0.00
30 - 44.9	78	β-Thalassaemia Major	20	25.64
		β-Thalassaemia Intermedia	12	15.38
		HbE/β-Thalassaemia	34	43.59
		HbH Disease	12	15.38
		Others	0	0.00
45 - 59.9	37	β-Thalassaemia Major	8	21.62
		β-Thalassaemia Intermedia	8	21.62
		HbE/β-Thalassaemia	15	40.54
		HbH Disease	6	16.22
		Others	0	0.00
Above 60	10	β-Thalassaemia Major	1	10.00
		β-Thalassaemia Intermedia	2	20.00
		HbE/β-Thalassaemia	1	10.00
		HbH Disease	6	60.00
		Others	0	0.00
Total			643	

4.2.2 Gender

As of 2019, there are 301 male patients (46.81%) and 342 female patients (53.19%) registered in Johor. The distribution of male and female patients in each hospital is as shown in Table 4.6.

Table 4.6: Distribution of Patients in Johor According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Sultanah Aminah	145	22.55	177	27.53
Hospital Sultan Ismail	48	7.47	45	7.00
Hospital Pakar Sultanah Fatimah	44	6.84	45	7.00
Hospital Enche' Besar Hajjah Kalsom	20	3.11	23	3.58
Hospital Segamat	15	2.33	21	3.27
Hospital Sultanah Nora Ismail	18	2.80	12	1.87
Hospital Mersing	11	1.71	19	2.95
Total	301	46.81	342	53.19

4.2.3 Ethnic Group

The distribution of patients in Johor according to major ethnic groups are 75.43% Malay (485 patients), 18.20% Chinese (117), and 1.56% Indian (10). Other ethnicities such as Orang Asli, Kadazan-Dusun, Pribumi Sabah and Sarawak contribute to 4.82% of patients (31). As shown in Figure 4.4, Malay patients contribute to the highest number of patients in each centre.

Figure 4.3: Distribution of Patients in Johor by Ethnic Group

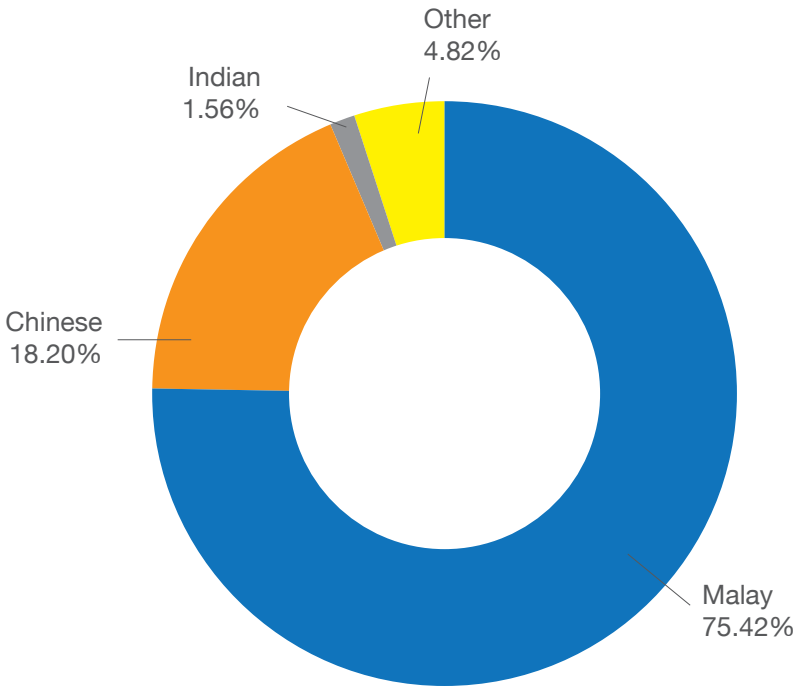
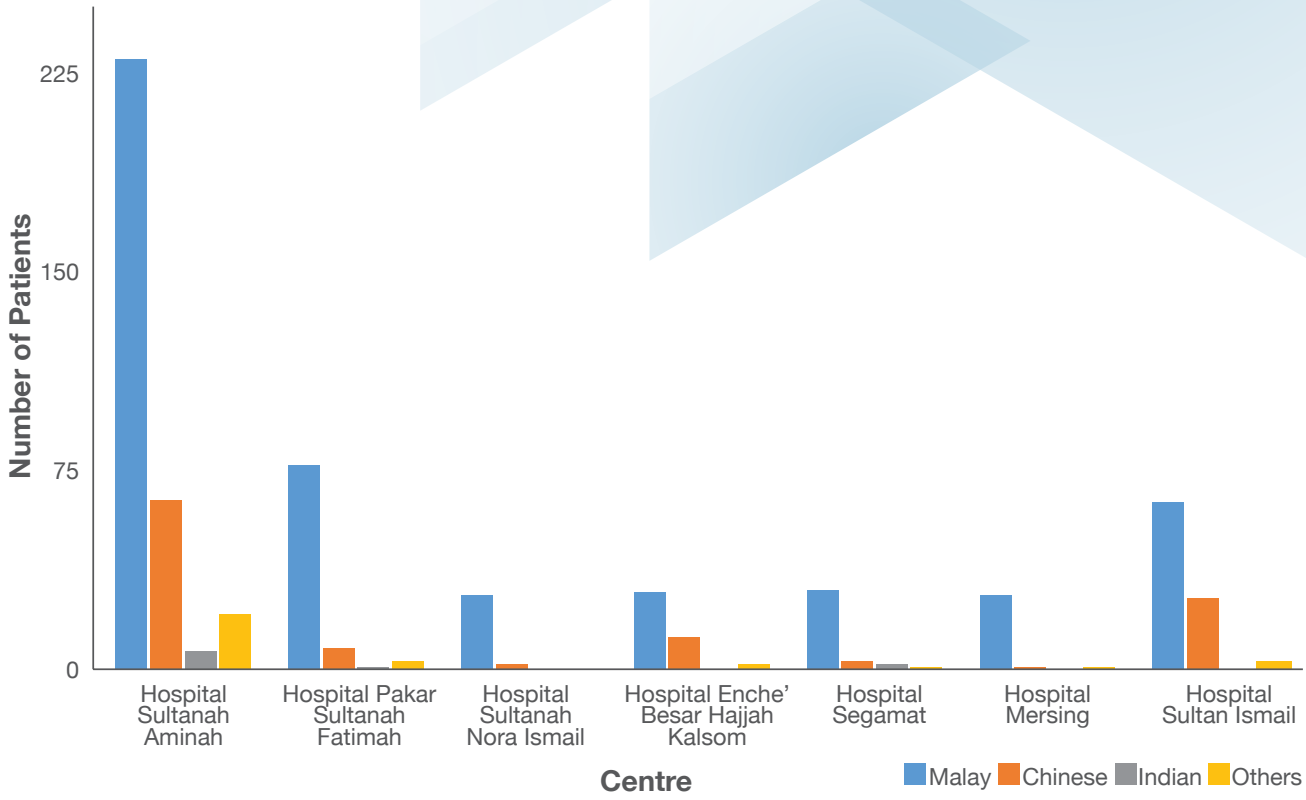


Figure 4.4: Distribution of Patients in Johor According to Ethnic Group by Centre



4.3. Diagnosis

Majority of the patients in Johor are afflicted by β-thalassaemia, as 291 patients have HbE/β-thalassaemia (45.26%), 199 have β-thalassaemia major (30.95%) and 42 have β-thalassaemia intermedia (6.53%). For α-thalassaemia, 109 patients have HbH disease (16.95%) and the remaining 2 patients have other types of α-thalassaemia (0.31%), as shown in Figure 4.5.

Figure 4.5: Distribution of Patients in Johor by Diagnosis

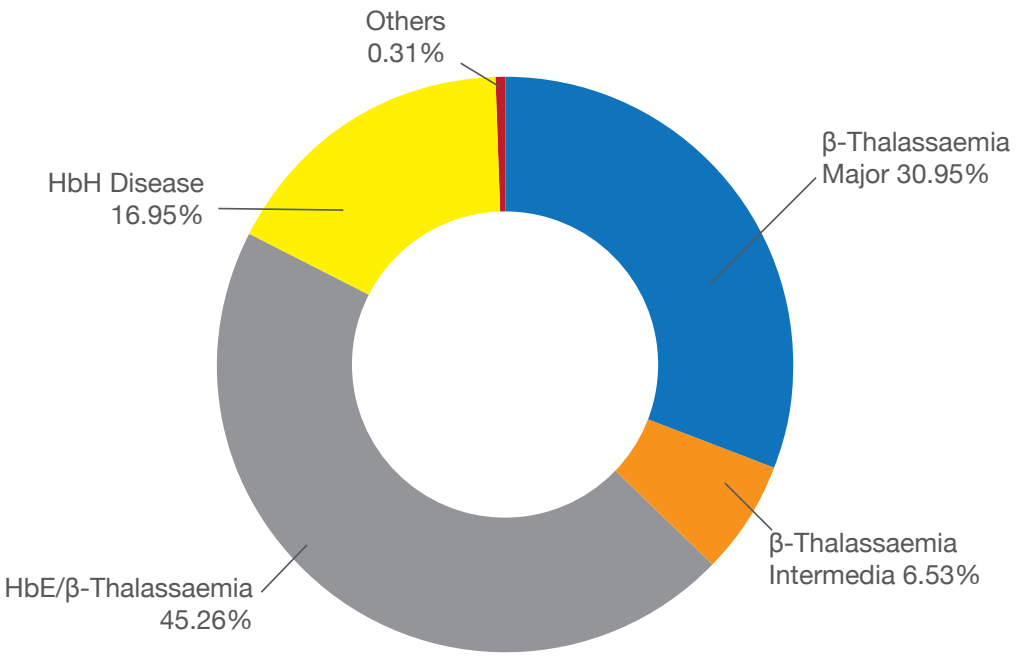


Table 4.7: Distribution of Patients in Johor According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number Of Patients (n)	Percentage (%)
β-Thalassaemia Major	199	Malay	124	19.28
		Chinese	51	7.93
		Indian	5	0.78
		Others	19	2.95
β-Thalassaemia Intermedia	42	Malay	29	4.51
		Chinese	7	1.09
		Indian	2	0.31
		Others	4	0.62
HbE/β-Thalassaemia	291	Malay	274	42.61
		Chinese	14	2.18
		Indian	0	0.00
		Others	3	0.47
HbH Disease	109	Malay	57	8.86
		Chinese	44	6.84
		Indian	3	0.47
		Others	5	0.78
Others	2	Malay	1	0.16
		Chinese	1	0.16
		Indian	0	0.00
		Others	0	0.00
Total			643	100.00

4.4 Treatment

4.4.1 Iron Chelation Therapy

A total of 428 patients are on iron chelation therapy in Johor, as shown in Table 4.8. A total of 314 patients (73.36%) are on monotherapy, in which the highest number of patients receive DFP, followed by DFX and DFO. Another 113 patients received a two-drug combination and only 1 patient is on a three-drug combination. The combination of DFO and DFP is the most common practice in a majority of the hospitals (Table 4.9). The youngest TDT patient who receives DFX is aged 3 years old. As shown in Table 4.10 and Figure 4.8, DFX is the drug of choice for the treatment of paediatric patients younger than 18 years old, while DFP is more widely used in adults.

Table 4.8: Distribution of Patients in Johor by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	50	11.68
DFP only	160	37.38
DFX only	104	24.30
DFO + DFP	90	21.03
DFP + DFX	8	1.87
DFO + DFX	15	3.50
DFO + DFP + DFX	1	0.23
Total	428	100.00

Table 4.9: Distribution of Patients in Johor According to Type of Iron Chelator Received by Centre

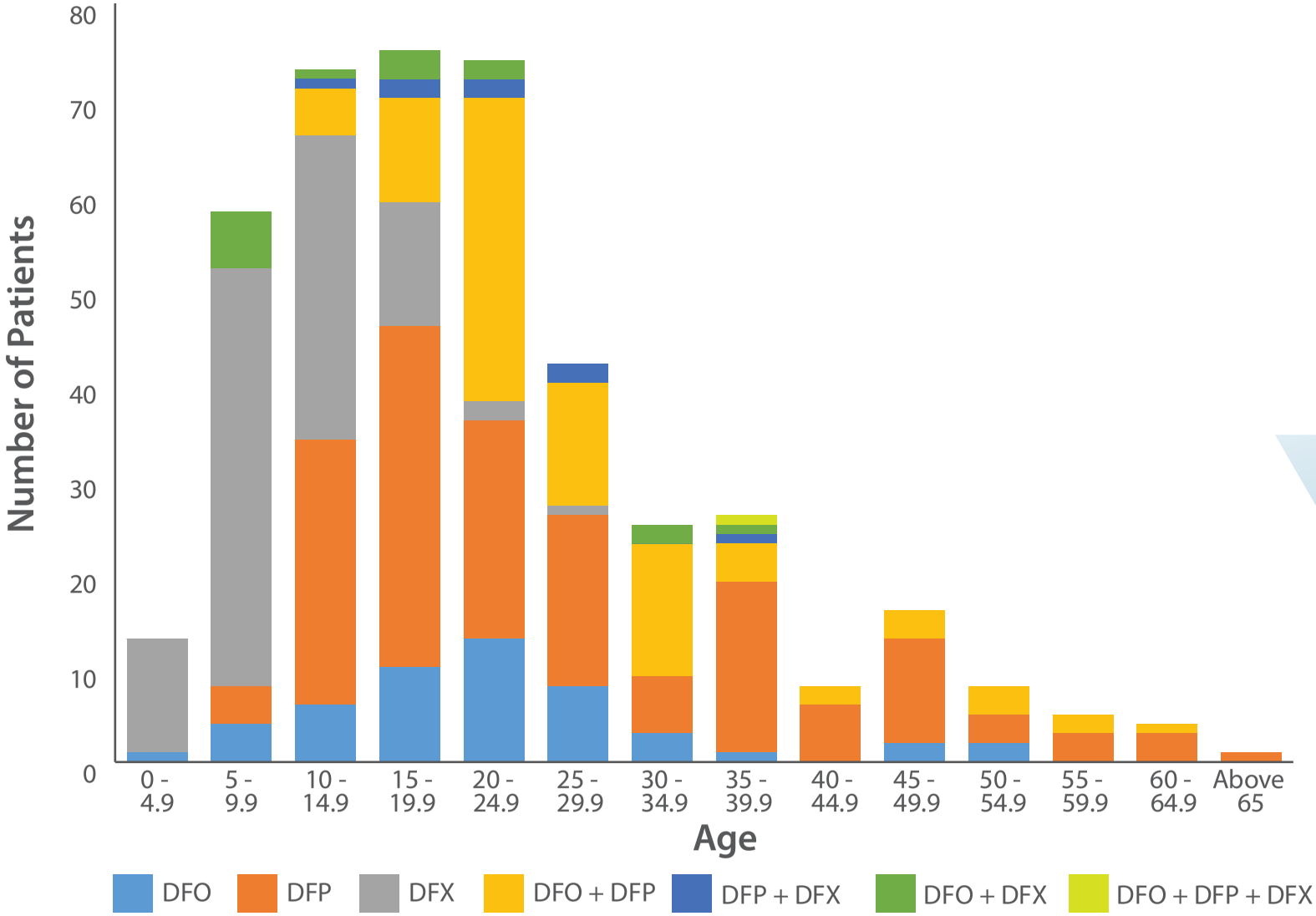
Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Sultanah Aminah	192	DFO only	22	5.14
		DFP only	88	20.56
		DFX only	22	5.14
		DFO + DFP	48	11.21
		DFP + DFX	6	1.40
		DFO + DFX	5	1.17
		DFO + DFP + DFX	1	0.23
Hospital Pakar Sultanah Fatimah	70	DFO only	13	3.04
		DFP only	27	6.31
		DFX only	12	2.80
		DFO + DFP	16	3.74
		DFP + DFX	1	0.23
		DFO + DFX	1	0.23
		DFO + DFP + DFX	0	0.00
Hospital Sultanah Nora Ismail	27	DFO only	5	1.17
		DFP only	3	0.70
		DFX only	12	2.80
		DFO + DFP	6	1.40
		DFP + DFX	0	0.00
		DFO + DFX	1	0.23
		DFO + DFP + DFX	0	0.00
Hospital Enche' Besar Hajjah Kalsom	39	DFO only	4	0.93
		DFP only	10	2.34
		DFX only	11	2.57
		DFO + DFP	12	2.80
		DFP + DFX	0	0.00
		DFO + DFX	2	0.47
		DFO + DFP + DFX	0	0.00
Hospital Segamat	26	DFO only	2	0.47
		DFP only	10	2.34
		DFX only	11	2.57
		DFO + DFP	1	0.23
		DFP + DFX	0	0.00
		DFO + DFX	2	0.47
		DFO + DFP + DFX	0	0.00
Hospital Mersing	20	DFO only	0	0.00
		DFP only	10	2.34
		DFX only	4	0.93
		DFO + DFP	6	1.40
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Sultan Ismail	54	DFO only	4	0.93
		DFP only	12	2.80
		DFX only	32	7.48
		DFO + DFP	1	0.23
		DFP + DFX	1	0.23
		DFO + DFX	4	0.93
		DFO + DFP + DFX	0	0.00
Total			428	100.00

Table 4.10: Distribution of Patients in Johor According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	144	DFO only	11	7.64
		DFP only	32	22.22
		DFX only	88	61.11
		DFO + DFP	5	3.47
		DFP + DFX	1	0.69
		DFO + DFX	7	4.86
		DFO + DFP + DFX	0	0.00
15 - 29.9	191	DFO only	31	16.23
		DFP only	77	40.31
		DFX only	16	8.38
		DFO + DFP	56	29.32
		DFP + DFX	6	3.14
		DFO + DFX	5	2.62
		DFO + DFP + DFX	0	0.00
30 - 44.9	59	DFO only	4	6.78
		DFP only	30	50.85
		DFX only	0	0.00
		DFO + DFP	20	33.90
		DFP + DFX	1	1.69
		DFO + DFX	3	5.08
		DFO + DFP + DFX	1	1.69
45 - 59.9	29	DFO only	4	13.79
		DFP only	17	58.62
		DFX only	0	0.00
		DFO + DFP	8	27.59
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	5	DFO only	0	0.00
		DFP only	4	80.00
		DFX only	0	0.00
		DFO + DFP	1	20.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			428	

Figure 4.8: Distribution of Patients in Johor According to Type of Iron Chelator Received by Age Group



4.4.2 Serum Ferritin Level

A total of 315 TDT patients in Johor have their serum ferritin level measured in 2019. More than half (51.75%) of these patients have a serum ferritin level lower than 2500 ng/mL. However, 55 patients (17.46%) have a severe iron overload with serum ferritin level above 5000 ng/mL (Table 4.11). Most patients with high ferritin levels are adolescents or adults who are not compliant to iron chelation therapy.

Table 4.11: Distribution of TDT Patients in Johor According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Aminah	139	26	8.25	55	17.46	35	11.11	20	6.35	3	0.95
Hospital Pakar Sultanah Fatimah	40	6	1.90	12	3.81	15	4.76	6	1.90	1	0.32
Hospital Sultanah Nora Ismail	18	0	0.00	4	1.27	10	3.17	4	1.27	0	0.00
Hospital Enche' Besar Hajjah Kalsom	25	3	0.95	8	2.54	10	3.17	4	1.27	0	0.00
Hospital Segamat	18	1	0.32	6	1.90	6	1.90	3	0.95	2	0.63
Hospital Mersing	20	3	0.95	3	0.95	6	1.90	6	1.90	2	0.6
Hospital Sultan Ismail	55	10	3.1	26	8.25	15	4.76	4	1.27	0	0.00
Total	315	49	15.56	114	36.19	97	30.79	47	14.92	8	2.54

4.5 Conclusion

There are 12 hospitals in Johor; however, most patients are in the Johor Bahru district due to the high population in the area and the presence of 2 main haematology centres, HSA and HSI. There are no patients at HTSMTI Kulaijaya, Hospital Pontian, Hospital Kota Tinggi and Hospital Tangkak, as these are district hospitals. There are 643 patients in Johor registered in the MTR in 2019. Of these, 562 patients are alive, 14 patients were cured by stem cell transplant and another 67 patients were lost to-follow-up. There were 3 deaths recorded, although these were not related to thalassaemia complications. Twelve new cases were recorded in 2019, of which 9 patients were born in 2018. Most of the new-birth patients are diagnosed as β-thalassaemia.

The youngest patient in Johor is an 11-month-old at HSI, whereas the oldest patient Is a 70-year-old under HSA follow-up. There were 301 male and 342 female patients from Johor registered in MTR in 2019. Majority of the thalassaemia patients in Johor are Malay, followed by Chinese, Indian and other ethnicities, i.e. Orang Asli, Kadazan-Dusun, and Pribumi Sabah & Sarawak.

Most patients (45.26%) in Johor are diagnosed with HbE/β-thalassaemia, followed by β-thalassaemia major (30.95%), HbH disease (16.95%), and β-thalassaemia intermedia (6.53%). The remaining 2 patients (0.31%) have other types of α-thalassaemia. A total of 428 patients are receiving iron chelation therapy. For the monotherapy, DFP has the highest number of patients (314), followed by DFX and DFO. Meanwhile, 113 patients receive a 2-drug combination (DFO + DFP is the most common regime prescribed) and only 1 patient receive a 3-drug combination of iron chelators.

There are 315 TDT patients with recorded serum ferritin level in 2019. Half of these patients (51.75%) have a serum ferritin level lower than 2500 ng/mL. Most patients with high serum ferritin level have either poor adherence to iron chelation therapy, a non-functioning DFO pump, experience severe side effects from drug or are unable to tolerate alternative iron chelators. This data needs to be updated, especially for the adult patients.

5 Kedah

5.1 Introduction

Kedah has an estimated population of 2,180,600 in 2019 (Department of Statistics Malaysia). Most thalassaemia patients in Kedah are receiving care at Hospital Sultanah Bahiyah, Hospital Sultan Abdul Halim and Hospital Kulim.

5.2 Patient Demographics

There are a total of 715 living patients in Kedah. Two patients were cured by stem cell transplant, 103 were lost to follow-up and 32 patients have succumbed to the disease. The distribution of patients in Kedah is shown in Table 5.1. There are no deaths reported among thalassaemia patients in 2019.

Table 5.1: Distribution of Patients in Kedah by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Sultanah Bahiyah, Alor Setar	359	50.21
Hospital Baling	5	0.70
Hospital Jitra	0	0.00
Hospital Kuala Nerang	3	0.42
Hospital Kulim	126	17.62
Hospital Langkawi	54	7.55
Hospital Sik	10	1.40
Hospital Sultan Abdul Halim, Sungai Petani	158	22.10
Hospital Yan	0	0.00
Total	715	100.00

Table 5.2: Distribution of Patients in Kedah by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	610
Cured by Stem Cell Therapy	2
Total	612
Lost to Follow-up	103
Total	715
Deaths in 2019	0
Cumulative Reported Deaths	32

Table 5.3: Cumulative Causes of Death in Kedah Since 2007

Causes of Death	Number of Patients	Percentage (%)
Infections	25	78.13
Cardiac Causes	1	3.13
Central Nervous System Event	1	3.13
Liver Disease	1	3.13
Motor Vehicle Accident	3	9.38
Others	1	3.13
Total	32	100.00

As shown in Table 5.3, there were 32 deaths of thalassaemia patients in Kedah since 2007. The table also shows the number of deaths according to each respective cause. The most common cause of death among thalassaemia patients was infection.

5.2.1 Age Group

A total of 443 patients (61.95%) in Kedah are between 0-19.9 years old. The youngest patient in Kedah is a 16-month-old and the oldest is 71 years old (β-thalassaemia intermedia).

Figure 5.1: Distribution of Patients in Kedah by Age Group

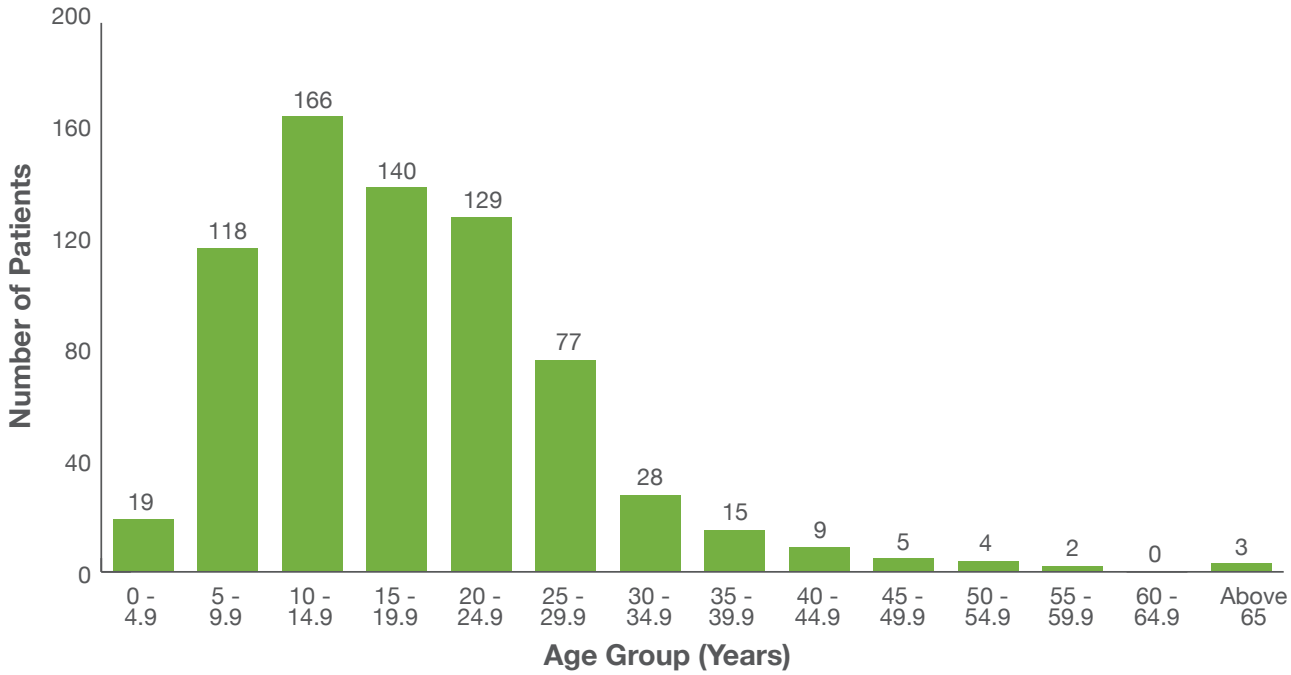


Table 5.4: Distribution of Patients in Kedah According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	303	β-Thalassaemia Major	38	12.54
		β-Thalassaemia Intermedia	26	8.58
		HbE/ β-Thalassaemia	125	41.25
		HbH Disease	92	30.36
		Others	22	7.26
15 - 29.9	346	β-Thalassaemia Major	59	17.05
		β-Thalassaemia Intermedia	18	5.20
		HbE/ β-Thalassaemia	155	44.80
		HbH Disease	89	25.72
		Others	25	7.23
30 - 44.9	52	β-Thalassaemia Major	11	21.15
		β-Thalassaemia Intermedia	6	11.54
		HbE/ β-Thalassaemia	20	38.46
		HbH Disease	13	25.00
		Others	2	3.85
45 - 59.9	11	β-Thalassaemia Major	1	9.09
		β-Thalassaemia Intermedia	4	36.36
		HbE/ β-Thalassaemia	1	9.09
		HbH Disease	4	36.36
		Others	1	9.09
60 and above	3	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	33.33
		HbE/ β-Thalassaemia	0	0.00
		HbH Disease	1	33.33
		Others	1	33.33
Total			715	

5.2.2 Gender

There are 379 male patients (53.02%) and 336 female patients (46.99%) in Kedah. The distribution of male and female patients in each district hospital is as shown in Table 5.5 and Figure 5.2.

Table 5.5: Distribution of Patients in Kedah According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Sultanah Bahiyah	178	24.90	181	25.31
Hospital Baling	3	0.42	2	0.28
Hospital Jitra	0	0.00	0	0.00
Hospital Kuala Nerang	0	0.00	3	0.42
Hospital Kulim	66	9.23	60	8.39
Hospital Langkawi	25	3.50	29	4.06
Hospital Sik	9	1.26	1	0.14
Hospital Sultan Abdul Halim	98	13.71	60	8.39
Hospital Yan	0	0.00	0	0.00
Total	379	53.02	336	46.99

5.2.3 Ethnic Group

The distribution of patients in Kedah by ethnic group is shown in Table 5.6. Majority of the patients (669 patients, 93.57%) are Malay, 20 patients (2.80%) are Chinese and 3 patients (0.42%) are of Indian descent. Another 23 patients (3.22%) are of other ethnicities. Figure 5.2 shows the distribution of patients in Kedah according to ethnic group by centre.

Figure 5.2: Distribution of Patients in Kedah by Ethnic Group

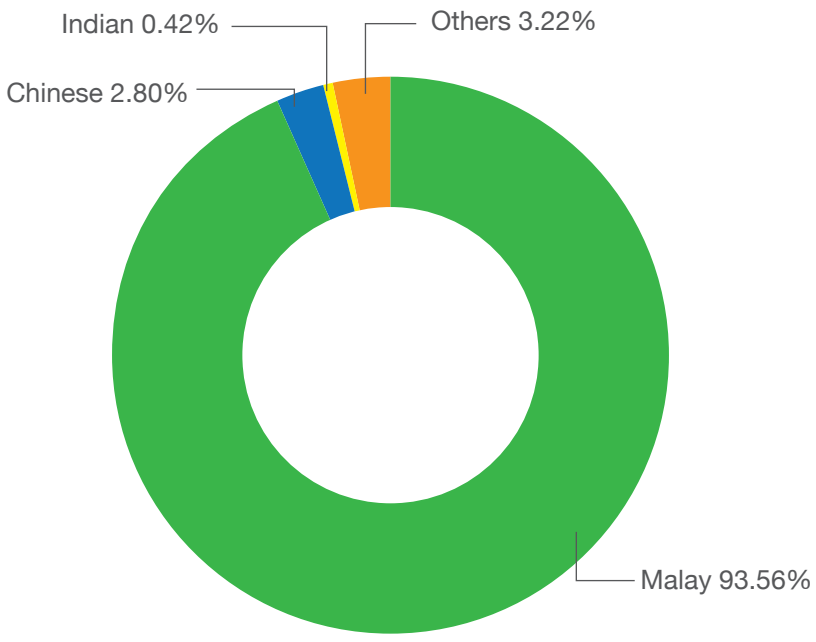


Table 5.6: Distribution of Patients in Kedah According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Thai		Mixed		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Bahiyah	331	46.29	10	1.40	1	0.14	15	2.10	0	0.00	2	0.28
Hospital Baling	5	0.70	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Jitra	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kuala Nerang	2	0.28	0	0.00	0	0.00	1	0.14	0	0.00	0	0.00
Hospital Kulim	122	17.06	2	0.28	1	0.14	0	0.00	0	0.00	1	0.14
Hospital Langkawi	52	7.27	2	0.28	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Sik	9	1.26	0	0.00	0	0.00	1	0.14	0	0.00	0	0.00
Hospital Sultan Abdul Halim	148	20.70	6	0.84	1	0.14	2	0.28	0	0.00	1	0.14
Hospital Yan	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	669	93.57	20	2.80	3	0.42	19	2.66	0	0.00	4	0.56

5.3 Diagnosis

HbE/β-thalassaemia is the most common type of thalassaemia in Kedah (301 patients, 42.10%), followed by HbH disease (199 patients, 27.83%), β-thalassaemia major (109 patients, 15.24%), and β-thalassaemia intermedia (55 patients, 7.69%). Fifty-one patients have other diagnoses (7.13%).

Figure 5.3: Distribution of Patients in Kedah by Diagnosis

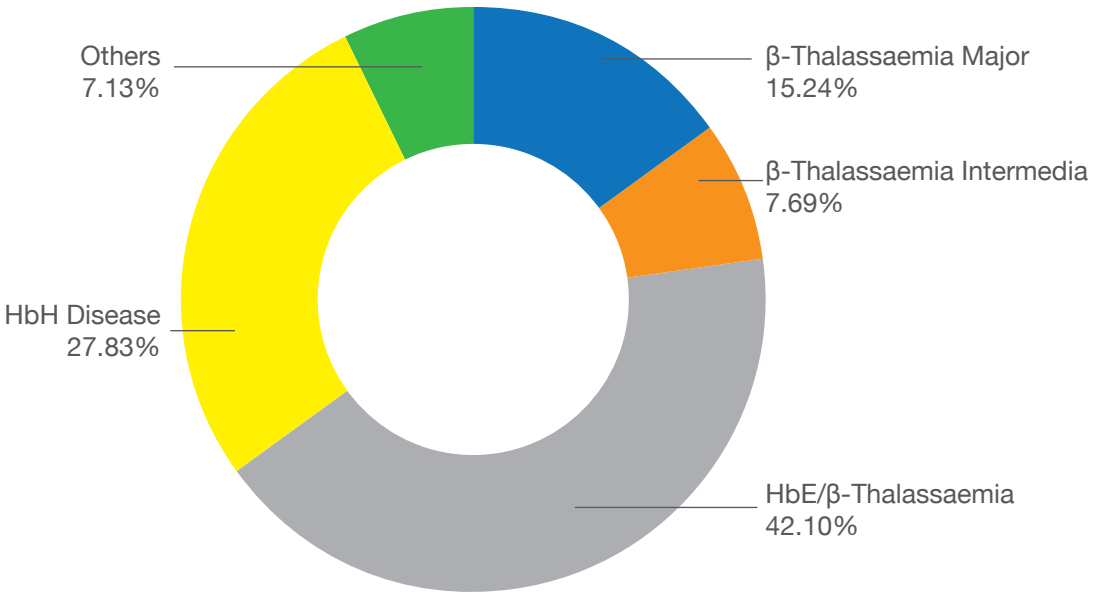


Table 5.7: Distribution of Patients in Kedah According to Diagnosis by Centre

Centre	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/ β-Thalassaemia		Hbh Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Bahiyah	67	9.37	22	3.08	146	20.42	107	14.97	17	2.38
Hospital Baling	3	0.42	0	0.00	1	0.14	1	0.14	0	0.00
Hospital Jitra	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kuala Nerang	0	0.00	0	0.00	1	0.14	0	0.00	2	0.28
Hospital Kulim	11	1.54	14	1.96	54	7.55	34	4.76	13	1.82
Hospital Langkawi	3	0.42	5	0.70	18	2.52	20	2.80	8	1.12
Hospital Sik	1	0.14	0	0.00	8	1.12	1	0.14	0	0.00
Hospital Sultan Abdul Halim	24	3.36	14	1.96	73	10.21	36	5.03	11	1.54
Hospital Yan	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	109	15.24	55	7.69	301	42.10	199	27.83	51	7.13

Table 5.8: Distribution of Patients in Kedah According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients	Percentage (%)
B-Thalassaemia Major	109	Malay	99	13.85
		Chinese	5	0.70
		Indian	1	0.14
		Others	4	0.56
B-Thalassaemia Intermedia	55	Malay	51	7.13
		Chinese	3	0.42
		Indian	0	0.00
		Others	1	0.14
Hbe/B-Thalassaemia	301	Malay	285	39.86
		Chinese	10	1.40
		Indian	2	0.28
		Others	4	0.56
Hbh Disease	199	Malay	184	25.73
		Chinese	2	0.28
		Indian	0	0.00
		Others	13	1.82
Others	51	Malay	50	6.99
		Chinese	0	0.00
		Indian	0	0.00
		Others	1	0.14
Total	715		715	100.00

5.4 Treatment

5.4.1 Iron Chelation Therapy

Of the 391 patients on chelation in Kedah, 51.66% are on oral DFX, 18.93% are on combination therapy, 9.46% are on subcutaneous DFO and 19.95% are on oral DFP.

Table 5.9: Distribution of Patients in Kedah by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	37	9.46
DFP only	78	19.95
DFX only	202	51.66
DFO + DFP	48	12.28
DFP + DFX	12	3.07
DFO + DFX	14	3.58
DFO + DFP + DFX	0	0.00
Total	391	100.00

Table 5.10: Distribution of Patients in Kedah According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients	Percentage (%)
Hospital Sultanah Bahiyah	197	DFO only	10	2.56
		DFP only	43	11.00
		DFX only	100	25.58
		DFO + DFP	23	5.88
		DFP + DFX	11	2.81
		DFO + DFX	10	2.56
		DFO + DFP + DFX	0	0.00
Hospital Baling	3	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	2	0.51
		DFO + DFP	1	0.26
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Jitra	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kuala Nerang	2	DFO only	0	0.00
		DFP only	1	0.26
		DFX only	1	0.26
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Kulim	69	DFO only	8	2.05
		DFP only	19	4.86
		DFX only	25	6.39
		DFO + DFP	14	3.58
		DFP + DFX	1	0.26
		DFO + DFX	2	0.51
		DFO + DFP + DFX	0	0.00
Hospital Langkawi	28	DFO only	0	0.00
		DFP only	6	1.53
		DFX only	19	4.86
		DFO + DFP	1	0.26
		DFP + DFX	0	0.00
		DFO + DFX	2	0.51
		DFO + DFP + DFX	0	0.00
Hospital Sik	6	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	6	1.53
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Sultan Abdul Halim	86	DFO only	19	4.86
		DFP only	9	2.30
		DFX only	48	12.28
		DFO + DFP	9	2.30
		DFP + DFX	0	0.00
		DFO + DFX	1	0.26
		DFO + DFP + DFX	0	0.00
Hospital Yan	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			391	100

Table 5.11: Distribution of Patients in Kedah According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	152	DFO only	4	2.63
		DFP only	11	7.24
		DFX only	129	84.87
		DFO + DFP	5	3.29
		DFP + DFX	1	0.66
		DFO + DFX	2	1.32
		DFO + DFP + DFX	0	0.00
15 - 29.9	199	DFO only	32	16.08
		DFP only	46	23.12
		DFX only	66	33.17
		DFO + DFP	34	17.09
		DFP + DFX	10	5.03
		DFO + DFX	11	5.53
		DFO + DFP + DFX	0	0.00
30 - 44.9	33	DFO only	0	0.00
		DFP only	19	57.58
		DFX only	3	9.09
		DFO + DFP	9	27.27
		DFP + DFX	0	0.00
		DFO + DFX	2	6.06
		DFO + DFP + DFX	0	0.00
45 - 59.9	6	DFO only	1	16.67
		DFP only	2	33.33
		DFX only	2	33.33
		DFO + DFP	0	0.00
		DFP + DFX	1	16.67
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	100.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			391	

5.4.2 Serum Ferritin Level

There are 36 patients (13.53%) with serum ferritin level lower than 1000 ng/mL, 97 patients (36.47%) with serum ferritin level between 1000-2499 ng/mL, 64 patients (26.89) with serum ferritin level between 2500-4999 ng/mL, and 41 patients (17.22%) with serum ferritin level above 5000 ng/mL.

Table 5.12: Distribution of Patients in Kedah According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Baling	2	0	0.00	0	0.00	1	0.42	1	0.42	0	0.00
Hospital Sultanah Bahiyah	127	21	8.82	44	18.49	34	14.29	21	8.82	7	2.94
Hospital Kuala Nerang	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kulim	36	4	1.68	19	7.98	9	3.78	4	1.68	0	0.00
Hospital Langkawi	21	1	0.42	4	1.68	9	3.78	3	1.26	0	0.00
Hospital Sik	0	0	0.00	0	0.00	0	0.00	0	0.00	4	1.68
Hospital Sultan Abdul Halim	52	10	4.20	30	12.61	11	4.62	1	0.42	0	0.00
Hospital Yan	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	238	36	15.13	97	40.76	64	26.89	30	12.61	11	4.62

5.5 Conclusion

Kedah has the third **highest** number of thalassaemia patients in Malaysia. The most common type of thalassaemia in Kedah is HbE/β-thalassaemia, with 301 patients (42.10%), followed by HbH disease (199 patients, 27.83%). The median age group is 10-14.9 years old. Approximately 65% of the thalassaemia patients in Kedah is aged below 20 years old. A total of 391 patients are receiving chelation therapy, and oral DFX is the most common chelator used (in 51.66% of patients on chelators). Around 56% of patients achieved serum ferritin level below 2500 ng/mL.

In Kedah, the only hospital which provides day care facilities is Hospital Sultan Abdul Halim. There was no mortality reported in Kedah in 2019. However, from the previous cumulative deaths reported, infection seems to be the main cause of death among thalassaemia patients. A total of 103 patients were lost to follow-up, while 1 patient was reported to have hepatitis C.

6 Kelantan

6.1 Introduction

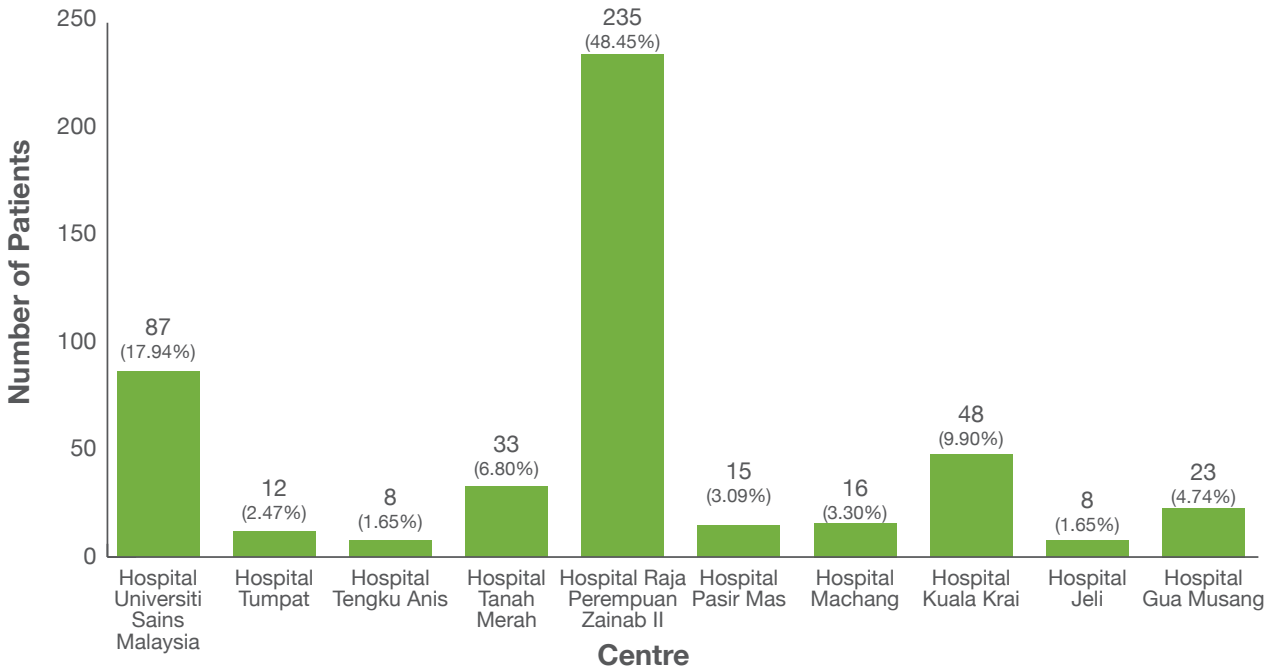
Kelantan is a state located in the northeast of Peninsular Malaysia. Kelantan's population is estimated at 1.86 million in 2019. The population is dominated by those of Malay descent, followed by the Chinese, Siamese and Indian. This state is managed by 10 administrative jurisdictions, namely Kota Bharu, Kubang Kerian, Tumpat, Pasir Puteh, Pasir Mas, Tanah Merah, Machang, Kuala Krai, Jeli, and Gua Musang.

There are 10 hospitals involved in providing treatment to thalassaemia patients in Kelantan. The total number of thalassaemia patients in Kelantan registered in the MTR are 485 patients (excluding patients who passed away). More than half of these patients received treatment at Hospital Raja Perempuan Zainab II (HRPZII) and Hospital Universiti Sains Malaysia (HUSM). HRPZII, HUSM and Hospital Kuala Krai are major specialist hospitals which provide day care services. Hospital Tanah Merah is a minor specialist hospital, whereas the remaining hospitals are non-specialist hospitals.

6.2 Patient Demographics

There are ten hospitals involved in providing treatment to thalassaemia patients in Kelantan. Figure 6.1 shows the distribution of thalassaemia patients by centre in Kelantan. HRPZII has the greatest number of thalassaemia patients, whereas Hospital Tengku Anis has the least.

Figure 6.1: Distribution of Patients in Kelantan by Centre



Data analysis was conducted on patients who are either living, lost to follow-up or cured by bone marrow transplant (485 patients in total). There are 256 TDT patients and 229 NTDT patients in Kelantan. Only 1 patient who was successfully cured by bone marrow transplant in Kelantan. No deaths of thalassaemia patients were recorded in 2019.

Table 6.1: Distribution of Patients in Kelantan by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	425
Cured by Stem Cell Therapy	1
Lost to Follow-up	59
Total	485
Deaths in 2019	0
Cumulative Reported Deaths	34

Table 6.2: Cumulative Causes of Death in Kelantan Since 2007

Causes of Death	Number of Patients
Infections	14
Cardiac Complications	15
Endocrine Complications	1
Thalassaemia	1
Thrombosis	1
Unknown	1
Others: Drowned	1
Total	34

There were 34 deaths of thalassaemia patients in Kelantan since 2007. Table 6.2 shows the causes of these deaths and the number of patients under each cause of death. The most common causes of death among thalassaemia patients were infections and cardiac causes. There was 1 death whereby the cause was not known, as the patient died at home without post-mortem performed.

6.2.1 Age Group

Majority (51.55%) of thalassaemia patients in Kelantan are aged between 10-25 years old, while 4 patients are above 65 years old.

Table 6.3: Distribution of Patients in Kelantan by Age Group

Age Group (Years)	Number of Patients (n)	Percentage (%)
0 – 4.9	22	4.54
5 – 9.9	63	12.99
10 – 14.9	89	18.35
15 – 19.9	72	14.85
20 – 24.9	89	18.35
25 – 29.9	56	11.55
30 – 34.9	40	8.25
35 – 39.9	15	3.09
40 – 44.9	9	1.86
45 – 49.9	15	3.09
50 – 54.9	6	1.24
55 – 59.9	3	0.62
60 – 64.9	2	0.41
Above 65	4	0.82
Total	485	100.00

Figure 6.2: Distribution of Patients in Kelantan by Age Group

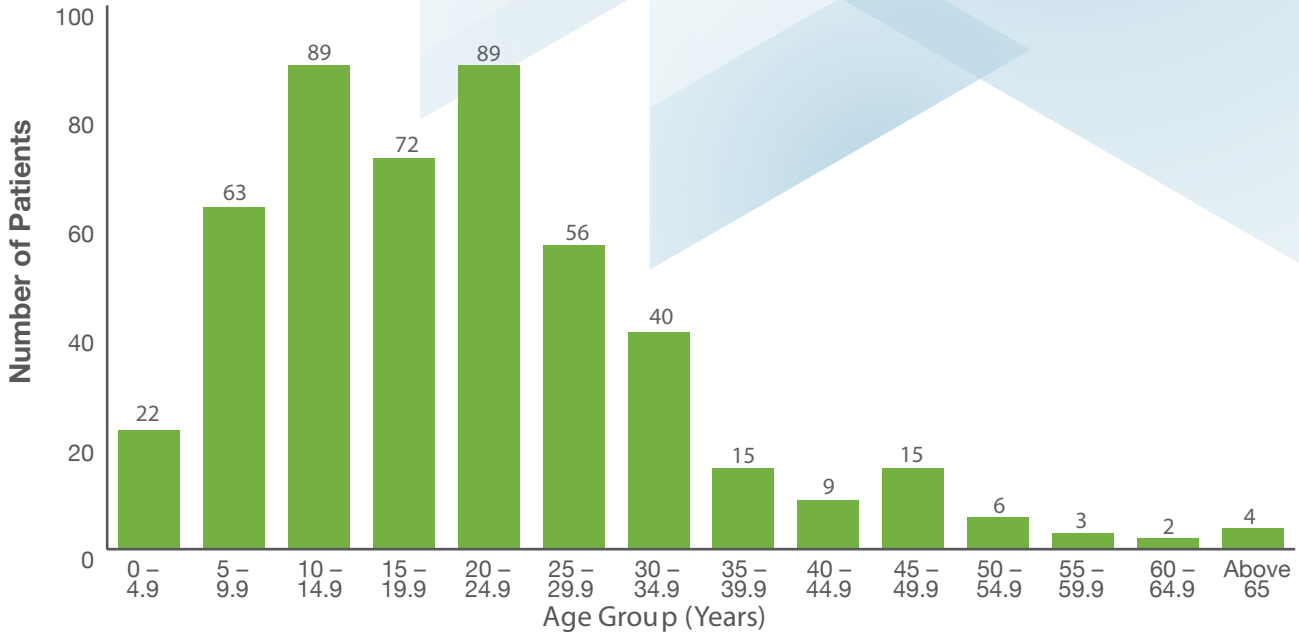


Table 6.4: Distribution of Patients in Kelantan According to Diagnosis by Age Group

Age (Years)	Total Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	174	β-Thalassaemia Major	14	8.05
		β-Thalassaemia Intermedia	77	44.25
		HbE/β-Thalassaemia	46	26.44
		HbH Disease	11	6.32
		Others	26	14.94
15 - 29.9	217	β-Thalassaemia Major	4	1.84
		β-Thalassaemia Intermedia	126	58.06
		HbE/β-Thalassaemia	55	25.35
		HbH Disease	4	1.84
		Others	28	12.90
30 - 44.9	64	β-Thalassaemia Major	5	7.81
		β-Thalassaemia Intermedia	34	53.13
		HbE/β-Thalassaemia	12	18.75
		HbH Disease	1	1.56
		Others	12	18.75
45 - 59.9	24	β-Thalassaemia Major	7	29.17
		β-Thalassaemia Intermedia	8	33.33
		HbE/β-Thalassaemia	6	25.00
		HbH Disease	3	12.50
		Others	0	0.00
60 and above	6	β-Thalassaemia Major	2	33.33
		β-Thalassaemia Intermedia	2	33.33
		HbE/β-Thalassaemia	1	16.67
		HbH Disease	1	16.67
		Others	0	0.00
Total			485	

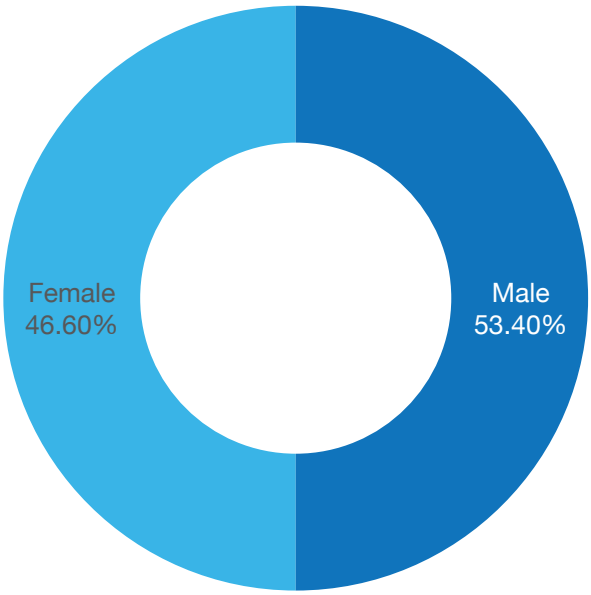
6.2.2 Gender

The distribution of male and female thalassaemia patients in Kelantan is almost equal.

Table 6.5: Distribution of Patients in Kelantan According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Raja Perempuan Zainab II	112	23.09	123	25.36
Hospital Pasir Mas	7	1.44	8	1.65
Hospital Tumpat	5	1.03	7	1.44
Hospital Machang	4	0.82	12	2.47
Hospital Tanah Merah	17	3.51	16	3.30
Hospital Tengku Anis, Pasir Puteh	7	1.44	1	0.21
Hospital Gua Musang	12	2.47	11	2.27
Hospital Kuala Krai	20	4.12	28	5.77
Hospital Universiti Sains Malaysia	36	7.42	51	10.52
Hospital Jeli	6	1.24	2	0.41
Total	226	46.60	259	53.40

Figure 6.3: Distribution of Patients in Kelantan by Gender



6.2.3 Ethnic Group

Thalassaemia patients in Kelantan consist of mainly Malay patients (94.85%), as depicted in Figure 6.4 and Table 6.6. This is because the Kelantanese population is dominated by the Malay ethnicity.

Table 6.6: Distribution of Patients in Kelantan by Ethnic Group

Ethnic Group		Number of Patients (n)	Percentage (%)
Malay		460	94.85
Chinese		14	2.89
Indian		0	0.00
Others			
a.	Thai	10	2.06
b.	Foreigner	1	0.21
Total		485	100.00

Figure 6.4: Distribution of Patients in Kelantan by Ethnic Group

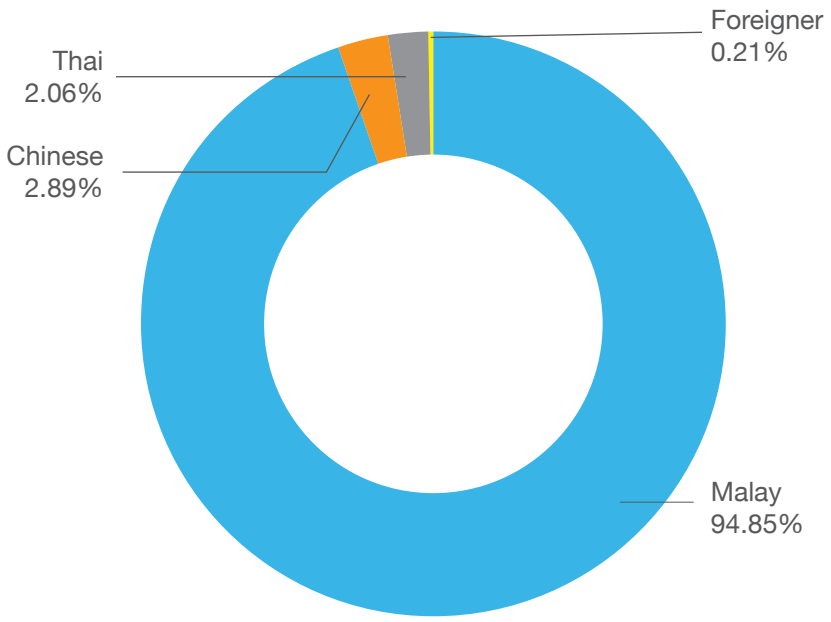


Table 6.7: Distribution of Patients in Kelantan According to Ethnic Group by Centre

Centre	Malay		Chinese		Thai		Foreigner	
	No.	%	No.	%	No.	%	No.	%
Hospital Raja Perempuan Zainab II	219	45.15	8	1.65	7	1.44	1	0.21
Hospital Pasir Mas	15	3.09	0	0.00	0	0.00	0	0.00
Hospital Tumpat	9	1.86	0	0.00	3	0.62	0	0.00
Hospital Machang	16	3.30	0	0.00	0	0.00	0	0.00
Hospital Tanah Merah	33	6.80	0	0.00	0	0.00	0	0.00
Hospital Tengku Anis, Pasir Puteh	8	1.65	0	0.00	0	0.00	0	0.00
Hospital Gua Musang	22	4.54	1	0.21	0	0.00	0	0.00
Hospital Kuala Krai	48	9.90	0	0.00	0	0.00	0	0.00
Hospital Universiti Sains Malaysia	82	16.91	5	1.03	0	0.00	0	0.00
Hospital Jeli	8	1.65	0	0.00	0	0.00	0	0.00
Total	460	94.85	14	2.89	10	2.06	1	0.21

6.3 Diagnosis

HbE/β-thalassaemia is the main diagnosis of thalassaemia patients in Kelantan, followed by HbH disease, β-thalassaemia major and β-thalassaemia intermedia. Twenty patients were diagnosed with thalassaemia types other than those mentioned above.

Table 6.8: Distribution of Patients in Kelantan by Diagnosis

Diagnosis	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	66	13.61
β-Thalassaemia Intermedia	32	6.60
HbE/β-Thalassaemia	247	50.93
HbH Disease	120	24.74
Others*	20	4.12
Total	485	100.00

**Other types of α-thalassaemia, excluding Hb Adana and HbH Constant Spring. Hb Adana and HbH Constant Spring are included under HbH disease.*

Figure 6.5: Distribution of Patients in Kelantan by Diagnosis

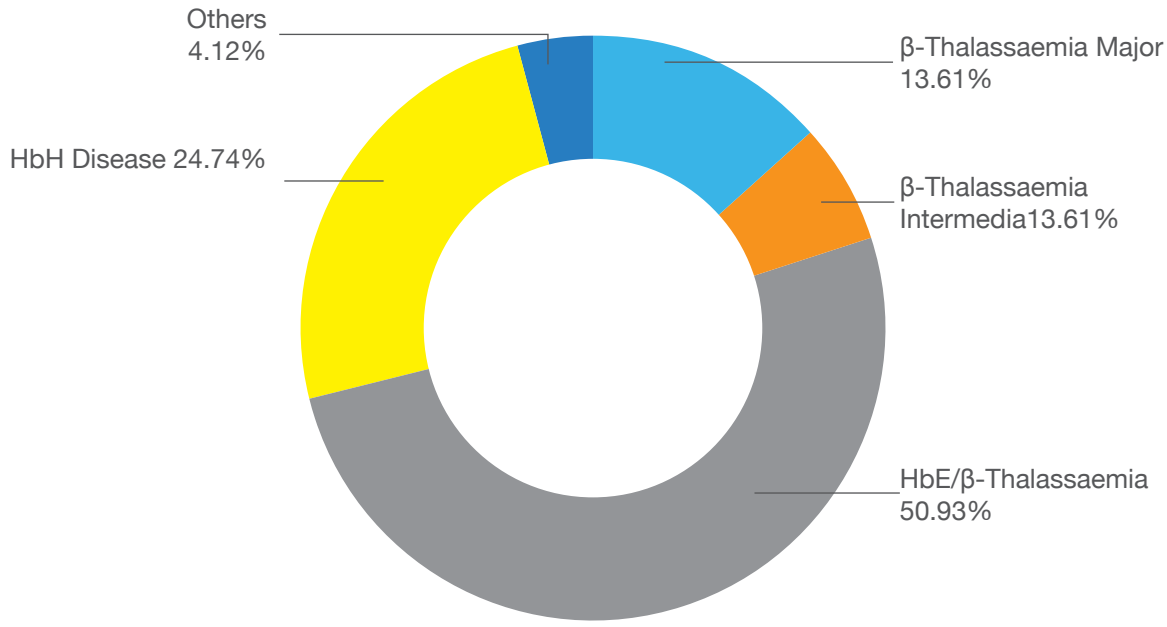


Table 6.9: Distribution of Patients in Kelantan According to Diagnosis by Centre

Centre	Diagnosis									
	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/ β-Thalassaemia		HbH Disease		Others*	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Raja Perempuan Zainab II	24	4.95	19	3.92	123	25.36	62	12.78	7	1.44
Hospital Pasir Mas	6	1.24	0	0.00	4	0.82	3	0.62	2	0.41
Hospital Tumpat	0	0.00	2	0.41	3	0.62	4	0.82	3	0.62
Hospital Machang	3	0.62	0	0.00	8	1.65	4	0.82	1	0.21
Hospital Tanah Merah	4	0.82	4	0.82	10	2.06	11	2.27	4	0.82
Hospital Tengku Anis, Pasir Puteh	2	0.41	0	0.00	5	1.03	1	0.21	0	0.00
Hospital Gua Musang	8	1.65	1	0.21	10	2.06	4	0.82	0	0.00
Hospital Kuala Krai	9	1.86	3	0.62	26	5.36	8	1.65	2	0.41
Hospital Universiti Sains Malaysia	8	1.65	3	0.62	54	11.13	22	4.54	0	0.00
Hospital Jeli	2	0.41	0	0.00	4	0.82	1	0.21	1	0.21
Total	**66**	**13.61**	**32**	**6.60**	**247**	**50.93**	**120**	**24.74**	**20**	**4.12**

Based on Table 6.9, HRPZ II has the highest number of thalassaemia patients in each type of diagnosis compared to other hospitals. This is due to HRPZ II being a tertiary hospital in Kelantan. Thus, most thalassaemia patients in Kelantan refer to HRPZ II for treatments.

Table 6.10: Distribution of Patients in Kelantan According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	66	Malay	62	12.78
		Chinese	2	0.41
		Thai	1	0.21
		Foreigner	1	0.21
β-Thalassaemia Intermedia	32	Malay	31	6.39
		Chinese	1	0.21
		Thai	0	0.00
		Foreigner	0	0.00
HbE/β--Thalassaemia	247	Malay	231	47.63
		Chinese	10	2.06
		Thai	6	1.24
		Foreigner	0	0.00
HbH Disease	120	Malay	117	24.12
		Chinese	0	0.00
		Thai	3	0.62
		Foreigner	0	0.00
Others	20	Malay	19	3.92
		Chinese	1	0.21
		Thai	0	0.00
		Foreigner	0	0.00
Total			**485**	**100.00**

Table 6.10 above showed that Malay patients form the majority in all 4 main types of thalassaemia in Kelantan.

6.4 Treatment

6.4.1 Iron Chelation Therapy

A total of 272 thalassaemia patients (56.08%) in Kelantan has been put on chelating agents. DFX is the most commonly prescribed agent (prescribed to 126 patients, 46.32%), followed by DFP (60 patients, 22.06%) and DFO (46 patients, 16.91%).

Table 6.11: Distribution of Patients in Kelantan by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	46	16.91
DFP only	60	22.06
DFX only	126	46.32
DFO + DFP	16	5.88
DFP + DFX	9	3.31
DFO + DFX	14	5.15
DFO + DFP + DFX	1	0.37
Total	272	100.00

Table 6.12: Distribution of Patients in Kelantan According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Raja Perempuan Zainab II	130	DFO only	11	4.04
		DFP only	35	12.87
		DFX only	65	23.90
		DFO + DFP	5	1.84
		DFP + DFX	5	1.84
		DFO + DFX	8	2.94
		DFO + DFP + DFX	1	0.37
Hospital Pasir Mas	6	DFO only	3	1.10
		DFP only	2	0.74
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	1	0.37
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tumpat	1	DFO only	0	0.00
		DFP only	1	0.37
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Machang	12	DFO only	3	1.10
		DFP only	1	0.37
		DFX only	7	2.57
		DFO + DFP	0	0.00
		DFP + DFX	1	0.37
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Tanah Merah	19	DFO only	0	0.00
		DFP only	4	1.47
		DFX only	15	5.51
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tengku Anis, Pasir Puteh	5	DFO only	3	1.10
		DFP only	0	0.00
		DFX only	2	0.74
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Gua Musang	16	DFO only	1	0.37
		DFP only	0	0.00
		DFX only	15	5.51
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kuala Krai	24	DFO only	3	1.10
		DFP only	3	1.10
		DFX only	15	5.51
		DFO + DFP	1	0.37
		DFP + DFX	1	0.37
		DFO + DFX	1	0.37
		DFO + DFP + DFX	0	0.00
Hospital Universiti Sains Malaysia	55	DFO only	21	7.72
		DFP only	13	4.78
		DFX only	5	1.84
		DFO + DFP	10	3.68
		DFP + DFX	1	0.37
		DFO + DFX	5	1.84
		DFO + DFP + DFX	0	0.00
Hospital Jeli	4	DFO only	1	0.37
		DFP only	1	0.37
		DFX only	2	0.74
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			272	100.00

Figure 6.6: Distribution of Patients in Kelantan by Type of Iron Chelator Received

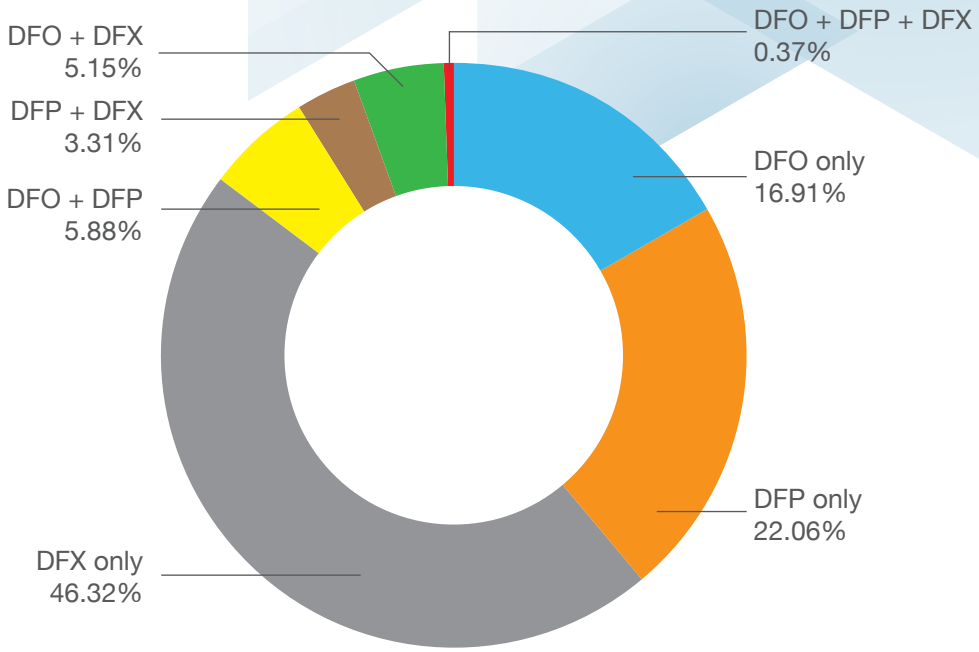


Table 6.13: Distribution of Patients in Kelantan According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	90	DFO only	8	8.89
		DFP only	9	10.00
		DFX only	66	73.33
		DFO + DFP	1	1.11
		DFP + DFX	1	1.11
		DFO + DFX	5	5.56
		DFO + DFP + DFX	0	0.00
15 - 29.9	133	DFO only	31	23.31
		DFP only	31	23.31
		DFX only	50	37.59
		DFO + DFP	8	6.02
		DFP + DFX	5	3.76
		DFO + DFX	7	5.26
		DFO + DFP + DFX	1	0.75
30 - 44.9	33	DFO only	5	15.15
		DFP only	12	36.36
		DFX only	8	24.24
		DFO + DFP	4	12.12
		DFP + DFX	2	6.06
		DFO + DFX	2	6.06
		DFO + DFP + DFX	0	0.00

45 - 59.9	13	DFO only	1	7.69
		DFP only	6	46.15
		DFX only	2	15.38
		DFO + DFP	3	23.08
		DFP + DFX	1	7.69
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	3	DFO only	1	33.33
		DFP only	2	66.67
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			272	100.00

6.4.2 Serum Ferritin Level

Out of 86 patients in Kelantan who have their serum ferritin levels regularly updated, 48 patients have a serum ferritin level lower than 2500 ng/mL.

Table 6.14: Distribution of Patients in Kelantan According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Raja Perempuan Zainab II	65	7	8.14	32	37.21	23	26.74	3	3.49	0	0.00
Hospital Pasir Mas	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Tumpat	2	1	1.16	1	1.16	0	0.00	0	0.00	0	0.00
Hospital Machang	7	0	0.00	2	2.33	3	3.49	2	2.33	0	0.00
Hospital Tanah Merah	1	0	0.00	0	0.00	1	1.16	0	0.00	0	0.00
Hospital Tengku Anis, Pasir Puteh	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Gua Musang	1	0	0.00	0	0.00	1	1.16	0	0.00	0	0.00
Hospital Kuala Krai	8	1	1.16	2	2.33	4	4.65	1	1.16	0	0.00
Hospital Universiti Sains Malaysia	1	0	0.00	1	1.16	0	0.00	0	0.00	0	0.00
Hospital Jeli	1	0	0.00	1	1.16	0	0.00	0	0.00	0	0.00
Total	86	9	10.47	39	45.35	32	37.21	6	6.98	0	0.00

6.5 Conclusion

The state of Kelantan has 485 thalassaemia patients. Most thalassaemia patients in Kelantan receive treatment at tertiary centres, i.e. HRPZII (235 patients out of 485, 48.45%) and HUSM (87 patients out of 485, 17.94%). No deaths of thalassaemia patients were recorded in the state in 2019.

Currently, only 1 patient is reported as cured by stem cell therapy. The distribution of male and female patients in Kelantan differs minimally, at 226 (46.60%) and 259 patients (53.40%), respectively. Most of the thalassaemia patients in Kelantan are of Malay descent (460 patients, 94.85%). HbE/β-thalassaemia contributes the highest number of patients in Kelantan (247 patients, 50.93%), followed by HbH disease (120 patients, 24.74%) and β-thalassaemia major (66 patients, 13.61%).

DFX is the most commonly prescribed chelating agent (126 patients, 46.32%). Eighty-six (33.59%) out of 256 TDT patients have their serum ferritin levels recorded. Of these, 48 patients have a serum ferritin level lower than 2500 ng/mL, while 38 patients have a serum ferritin level higher than 2500 ng/mL.

7 Melaka

7.1 Introduction

Melaka, dubbed The Historical State (or Negeri Bersejarah amongst locals), is the third smallest state in Malaysia, after Perlis and Penang. It is located in the southern region of the Malay Peninsula, by the Straits of Malacca. It borders Negeri Sembilan to the north and the state of Johor to the south. There are 3 districts in Melaka; Melaka Tengah, Alor Gajah and Jasin. The capital is Melaka Town. This historical city centre has been listed as a UNESCO World Heritage Site since 7 July 2008.

Melaka has a population of 930,700 as of 2019, comprising of Bumiputera (609,600, 65.50%), Chinese (including the Peranakan community; 217,500, 23.37%), Indian (including the Chitty community; 51,400, 5.52%), other races (5,600, 0.60%), and non-Malaysian citizens (46,600, 5.01%) (Department of Statistics Malaysia).

There are three government hospitals in the state of Melaka, but no thalassaemia cases are reported from Hospital Alor Gajah and Hospital Jasin. Both Hospital Alor Gajah and Hospital Jasin are without specialist, and thalassaemia patients are referred to and managed by Hospital Melaka.

7.2 Patient Demographics

The are 246 living thalassaemia patients in Melaka. The cumulative number of deceased patients up to 2019 stands at 5 patients. There were 2 deaths in 2019.

Table 7.1: Distribution of Patients in Melaka by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Alor Gajah	0	0.00
Hospital Jasin	0	0.00
Hospital Melaka	246	100.00
Total	246	100.00

Table 7.2: Distribution of Patients in Melaka by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	214
Cured by Stem Cell Therapy	5
Total	219
Lost to Follow-up	27
Total	246
Deaths in 2019	2
Cumulative Reported Deaths	5

Table 7.3: Cumulative Causes of Death in Melaka Since 2007

Causes of Death	Number of Patients	Percentage (%)
Cardiac Complications	1	20.00
Renal Complications	1	20.00
Infections	2	40.00
Other	1	20.00
Total	5	100.00

7.2.1 Age Group

The youngest patient in Melaka is a year old and the eldest is 65 years old. Most patients fall within the 10-14.9 years old age group (50 patients, 20.33%). As shown in Table 7.4, HbE/β-thalassaemia is the most common diagnosis for patients below 60 years old. The 2 most senior patients in Melaka are diagnosed with β-thalassaemia intermedia and HbH disease.

Figure 7.1: Distribution of Patients in Melaka by Age Group

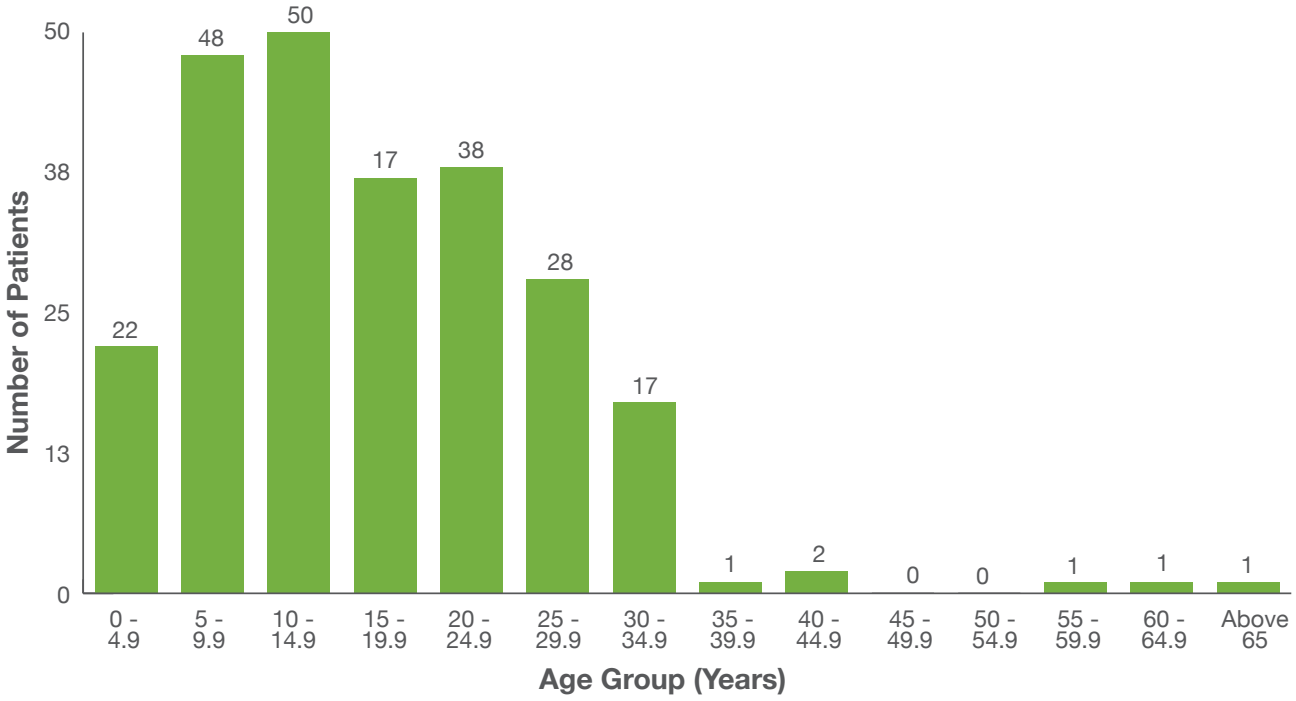


Table 7.4: Distribution of Patients in Melaka According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	120	β-Thalassaemia Major	26	21.67
		β-Thalassaemia Intermedia	2	1.67
		HbE/β-Thalassaemia	42	35.00
		HbH Disease	40	33.33
		Others	10	8.33
15 - 29.9	103	β-Thalassaemia Major	25	24.27
		β-Thalassaemia Intermedia	4	3.88
		HbE/β-Thalassaemia	52	50.49
		HbH Disease	18	17.48
		Others	4	3.88
30 - 44.9	20	β-Thalassaemia Major	5	25.00
		β-Thalassaemia Intermedia	2	10.00
		HbE/β-Thalassaemia	7	35.00
		HbH Disease	6	30.00
		Others	0	0.00
45 - 59.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	100.00
		HbH Disease	0	0.00
		Others	0	0.00
60 and above	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	50.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	50.00
		Others	0	0.00
Total			246	

7.2.2 Gender

Table 7.5 shows the distribution of thalassaemia patients in Melaka by gender. There are 124 (50.41%) male patients and 122 (49.59%) female patients in Melaka.

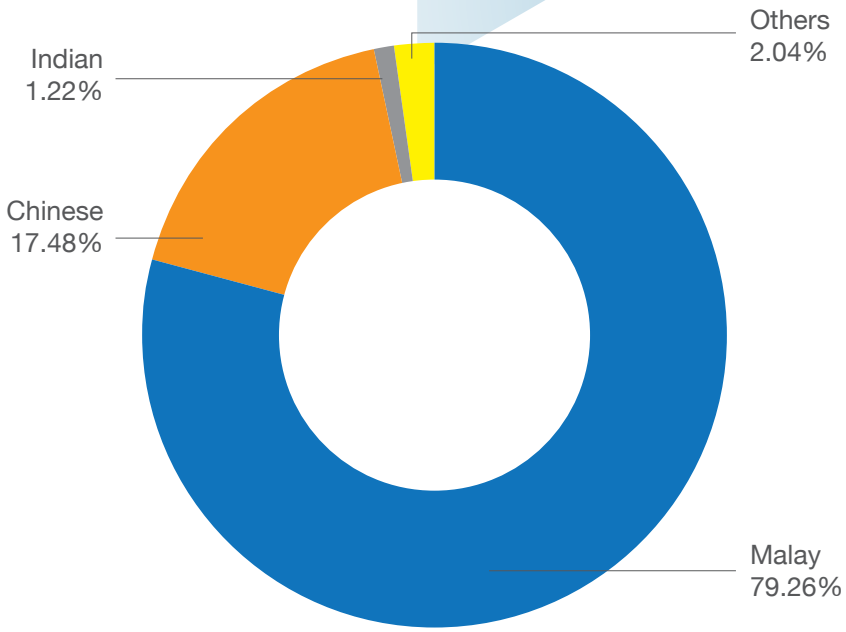
Table 7.5: Distribution of Patients in Melaka According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Alor Gajah	0	0	0	0
Hospital Jasin	0	0	0	0
Hospital Melaka	124	50.41	122	49.5
Total	124	50.41	122	49.59

7.2.3 Ethnic Group

Thalassaemia patients in Melaka are mostly Malay (195 patients, 79.27%), followed by Chinese (43 patients, 17.48%) and Indian (3 patients, 1.22%). Patients of other ethnicities include Bajau (1 patient, 0.41%), Iban (1 patient, 0.41%), Thai (2 patients, 0.88%) and Pribumi Sabah (1 patient, 0.41%).

Figure 7.2: Distribution of Patients in Melaka by Ethnic Group



7.3 Diagnosis

As shown in Figure 7.3, HbE/β-thalassaemia is the main diagnosis in Melaka with 102 patients (41.46%), followed by HbH disease with 65 patients (26.42%), β-thalassaemia major with 56 patients (22.76%), and β-thalassaemia intermedia with only 9 patients (3.66%). Malay patients are most commonly diagnosed with HbE/β-thalassaemia, whereas a majority of Chinese patients are diagnosed with β-thalassaemia major (Table 7.6).

Figure 7.3: Distribution of Patients in Melaka by Diagnosis

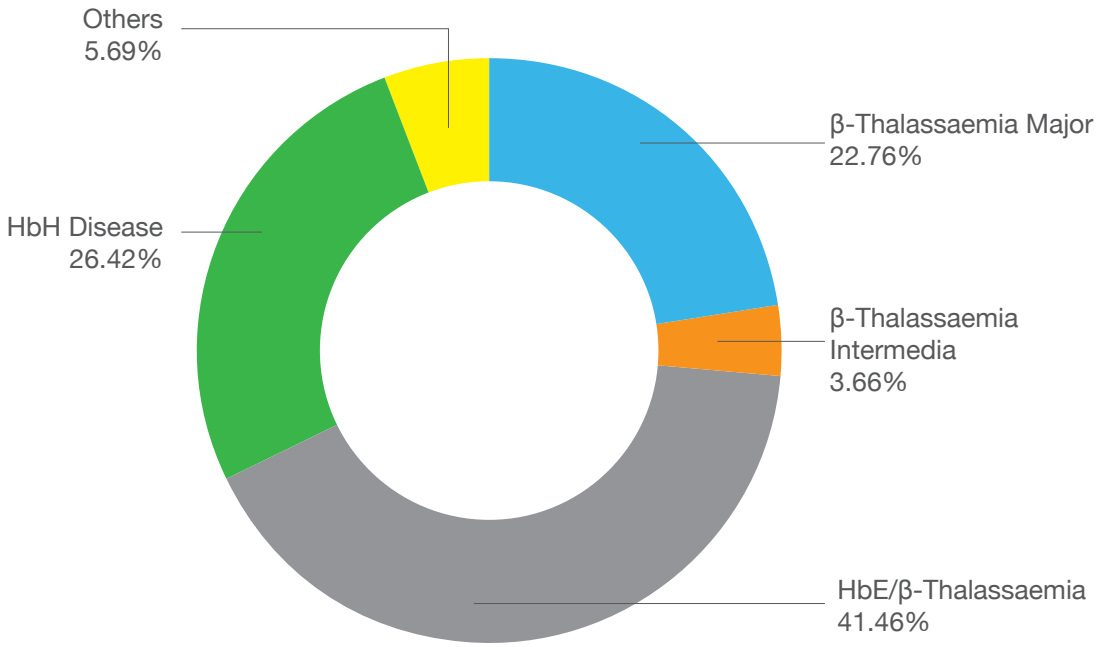


Table 7.6: Distribution of Patients in Melaka According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	56	Malay	34	13.82
		Chinese	22	8.94
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
β-Thalassaemia Intermedia	9	Malay	5	2.03
		Chinese	3	1.22
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	1	0.41
HbE/β-Thalassaemia	102	Malay	96	39.02
		Chinese	3	1.22
		Indian	2	0.81
		Kadazan-Dusun	0	0.00
		Others	1	0.41
HbH Disease	65	Malay	47	19.11
		Chinese	15	6.10
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	3	1.22
Others	14	Malay	13	5.28
		Chinese	0	0.00
		Indian	1	0.41
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Total			246	100.00

7.4 Treatment

7.4.1 Iron Chelation Therapy

Table 7.7 shows that there are 127 patients receiving iron chelators in Melaka (51.63%). Of these, 33 patients (25.98%) are on DFO, 54 patients (42.52%) are on DFX, 16 patients (12.60%) are on DFP and 24 patients (18.90%) receive a combination of 2 iron chelators. The youngest patient in Melaka receiving iron chelation therapy is 4 years old. As shown in Table 7.8, the youngest cohort of patients (aged 0-14.9 years) mostly receive DFX. Patients aged 15 to 60 years old who are on iron chelators commonly receive DFO therapy. Of the 3 patients above 60 years old, 2 of them are given DFP and 1 is on a combination of DFO and DFP.

Table 7.7: Distribution of Patients in Melaka by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	33	25.98
DFP only	16	12.60
DFX only	54	42.52
DFO + DFP	15	11.81
DFP + DFX	2	1.57
DFO + DFX	7	5.51
DFO + DFP + DFX	0	0.00
Total	127	100.00

Table 7.8: Distribution of Patients in Melaka According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	45	DFO only	5	11.11
		DFP only	0	0.00
		DFX only	36	80.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	4	8.89
		DFO + DFP + DFX	0	0.00
15 - 29.9	64	DFO only	21	32.81
		DFP only	7	10.94
		DFX only	18	28.13
		DFO + DFP	15	23.44
		DFP + DFX	2	3.13
		DFO + DFX	1	1.56
		DFO + DFP + DFX	0	0.00
30 - 44.9	15	DFO only	7	46.67
		DFP only	6	40.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	2	13.33
		DFO + DFP + DFX	0	0.00

45 - 59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	3	DFO only	0	0.00
		DFP only	2	66.67
		DFX only	0	0.00
		DFO + DFP	1	33.33
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			127	

7.4.2 Serum Ferritin Level

There are 71 patients in Melaka receiving regular transfusion who had their serum ferritin level measured. According to Table 7.9, there are 4 patients with serum ferritin level more than 10,000 ng/mL. The highest serum ferritin level measured is 18,985.80 ng/mL (19-year-old patient with diagnosed with HbE/β-thalassaemia). About half of TDT patients (50.70%) have serum ferritin levels lower than 2499 ng/mL.

Table 7.9: Distribution of Patients in Melaka According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Alor Gajah	0	0	0.00	0	0	0	0.00	0	0.00	0	0.00
Hospital Jasin	0	0	0.00	0	0	0	0.00	0	0.00	0	0.00
Hospital Melaka	71	12	16.90	24	33.80	16	22.54	15	21.13	4	5.63
Total	71	12	16.90	24	33.80	16	22.54	15	21.13	4	5.63

7.5 Conclusion

There are 246 living patients currently registered at Hospital Melaka. Of these, 214 are active patients, 5 patients are cured by stem cell therapy and 27 patients are lost to follow-up. No new thalassaemia births were recorded in 2019, whereas 2 patients were born in 2018.

Most patients in Melaka are aged between 10-14.9 years old. The patients' age ranged from a year old to 66 years old. Of the 246 thalassaemia patients in Melaka, 120 patients (48.78%) are under 15 years old and fall under paediatric category. A narrow majority of the patients are male, with a female to male ratio of 1:1.5. Malay patients constitute 79.27% of the total number of patients, followed by Chinese (17.48%), Indian (1.22%), Thai (0.81%), Bajau, Iban and Pribumi Sabah (each at 0.41%).

Most of the TDT patients in have HbE/β-thalassaemia (41.46%), followed by β-thalassaemia major (22.76%). On the other hand, most NTDT patients have HbH disease (26.42%), followed by β-thalassaemia intermedia (3.66%). Other diagnoses such as α-thalassaemia, δβ-thalassaemia and HbS with β-thalassaemia form 5.69% of the total patients in Melaka. The oldest surviving β-thalassaemia major patient is in the age range of 40-44.9 years old.

Despite of higher number of patients on iron chelation therapy and monitoring, only 54.20% of the patients receiving regular transfusion have their serum ferritin level measured. Of these, only 50.70% have a serum ferritin level below 2499 ng/mL. Overall, there is still a need to continue to educate, encourage and provide information to our patients to improve their iron status and their overall clinical outcome.

8 Negeri Sembilan

8.1 Introduction

Negeri Sembilan is located on the western coast of Peninsular Malaysia, just south of Kuala Lumpur. The state is bordered at the north by Selangor, east by Pahang and at the south by Melaka and Johor.

Negeri Sembilan has a collective population of 1.13 million (Department of Statistic Malaysia, 2019). This state consists of seven districts, namely Seremban, Port Dickson, Kuala Pilah, Jempol, Jelebu, Rembau and Tampin.

There are 6 government hospitals in the state of Negeri Sembilan, namely Hospital Tuanku Ja'afar (HTJ, Seremban), Hospital Tuanku Ampuan Najihah (HTAN, Kuala Pilah), Hospital Jelebu, Hospital Port Dickson, Hospital Jempol and Hospital Tampin.

The total number of thalassaemia patients registered in the MTR till 31st October 2019 in Negeri Sembilan is 202. Most of the patients (158 patients) receive their treatment at HTJS, followed by HTAN. HTJS is the only hospital in Negeri Sembilan which provides day care services, including blood transfusions for thalassaemia patients.

8.2 Patient Demographics

Patients are categorised as living, lost to follow-up or cured by transplant. The total number of living patients is 202, and the cumulative number of deceased patients as per 2019 is 10. No new birth was reported since 2018.

Table 8.1: Distribution of Patients in Negeri Sembilan by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Jelebu	6	2.97
Hospital Tuanku Ampuan Najihah, Kuala Pilah	35	17.33
Hospital Port Dickson	3	1.49
Hospital Tuanku Ja'afar, Seremban	158	78.22
Hospital Tampin	0	0.00
Total	202	100.00

Table 8.2: Distribution of Patients in Negeri Sembilan by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	170
Cured by Stem Cell Therapy	10
Total	180
Lost to Follow-up	22
Total	202
Deaths in 2019	1
Cumulative Reported Deaths	10

Table 8.3: Cumulative Causes of Death in Negeri Sembilan Since 2007

Causes of Death	Number of Patients	Percentage (%)
Infections	6	60.00
Cardiac Causes	2	20.00
Renal Failure	1	10.00
Liver Disease	1	10.00
Total	10	100.00

8.2.1 Age Group

The youngest patient in Negeri Sembilan is 2 years old, while the oldest is 66 years old. The age group between 20-24.9 years old has the highest number of patients (Figure 8.1). Table 8.4 shows that patients aged below 45 years are most commonly diagnosed with HbE/β-thalassaemia. There is 1 β-thalassaemia major patient aged above 60 years old.

Figure 8.1: Distribution of Patients in Negeri Sembilan by Age Group

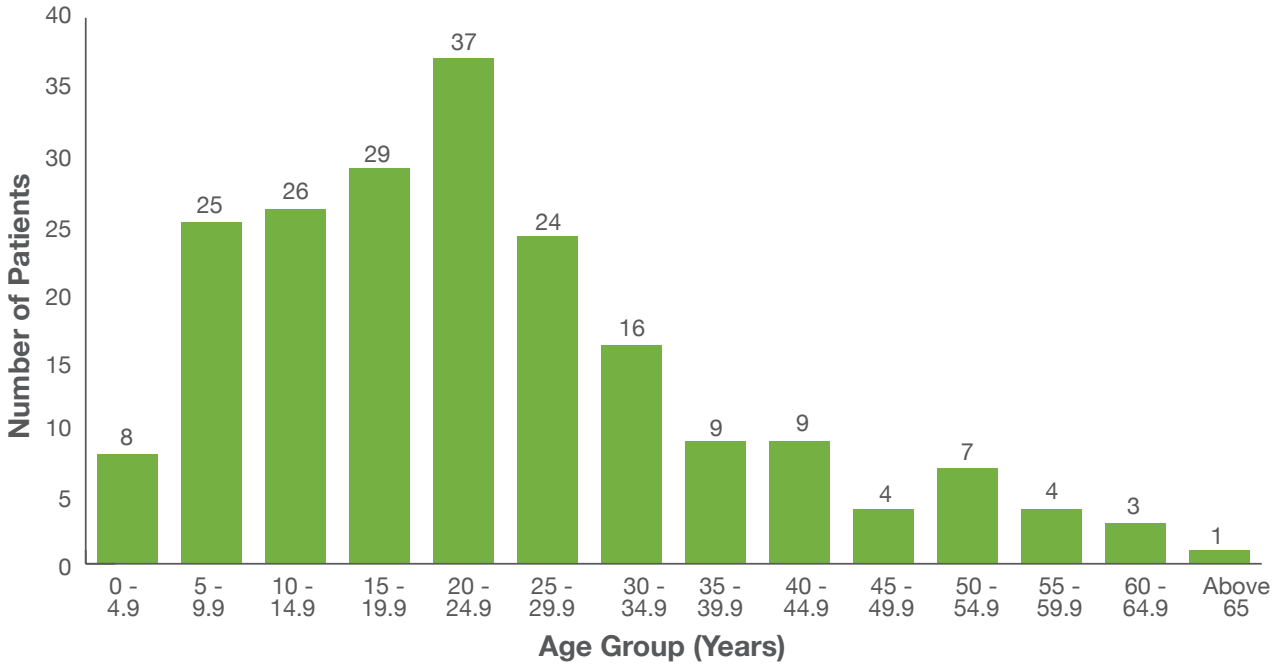


Table 8.4: Distribution of Patients in Negeri Sembilan According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	59	β-Thalassaemia Major	17	28.81
		β-Thalassaemia Intermedia	4	6.78
		HbE/β-Thalassaemia	23	38.98
		HbH Disease	8	13.56
		Others	7	11.86
15 - 29.9	90	β-Thalassaemia Major	28	31.11
		β-Thalassaemia Intermedia	3	3.33
		HbE/β-Thalassaemia	37	41.11
		HbH Disease	13	14.44
		Others	9	10.00
30 - 44.9	34	β-Thalassaemia Major	8	23.53
		β-Thalassaemia Intermedia	7	20.59
		HbE/β-Thalassaemia	11	32.35
		HbH Disease	5	14.71
		Others	3	8.82
45 - 59.9	15	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	8	53.33
		HbE/β-Thalassaemia	3	20.00
		HbH Disease	4	26.67
		Others	0	0.00
60 and above	4	β-Thalassaemia Major	1	25.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	2	50.00
		HbH Disease	1	25.00
		Others	0	0.00
Total			202	

8.2.2 Gender

Table 8.5 shows the distribution of patients in Negeri Sembilan by gender. Eighty-nine patients (44.06%) in Negeri Sembilan are male and 113 patients (55.94%) are female.

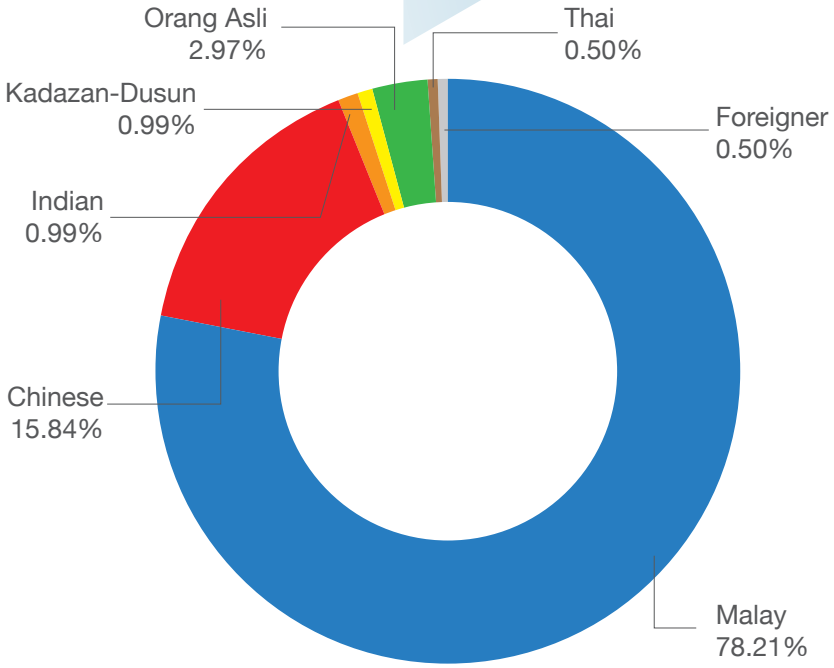
Table 8.5: Distribution of Patients in Negeri Sembilan According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Jelebu	3	1.49	3	1.49
Hospital Tuanku Ampuan Najihah, Kuala Pilah	25	12.38	10	4.95
Hospital Port Dickson	1	0.50	2	0.99
Hospital Tuanku Ja'a	60	29.70	98	48.51
Hospital Tampin	0	0.00	0	0.00
Total	89	44.06	113	55.94

8.2.3 Ethnic Group

The distribution of thalassaemia patients in Negeri Sembilan according to ethnic group are as follows: 158 out of 202 patients (78.21%) are Malay, 32 patients (15.84%) are Chinese, 2 patients (0.99%) are Indian, 2 patients (0.99%) are Kadazan-Dusun, 6 patients (2.97%) are Orang Asli, 1 patient (0.50%) is Thai and another 1 patient (0.50%) is Indonesian.

Figure 8.2: Distribution of Patients in Negeri Sembilan by Ethnic Group



8.3 Diagnosis

HbE/β-thalassaemia forms the main diagnosis with 76 patients. This is followed by β-thalassaemia major (54 patients), HbH disease (31 patients) and β-thalassaemia intermedia (22 patients). Nineteen patients have other diagnoses such as HbS disease and other types of α-thalassaemia (Figure 8.3). As shown in Table 8.6, the majority of HbE/β-thalassaemia patients are Malay (69 patients). Similarly, Malay patients also contribute the highest number of patients for other types of thalassaemia diagnoses.

Figure 8.3: Distribution of Patients in Negeri Sembilan by Diagnosis

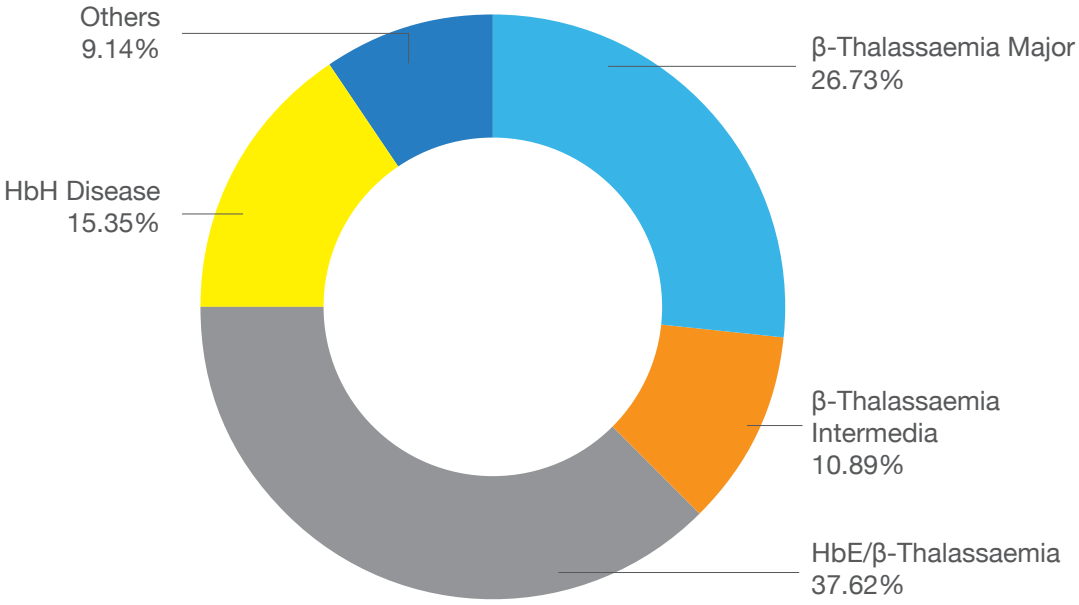


Table 8.6: Distribution of Patients in Negeri Sembilan According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	54	Malay	35	17.33
		Chinese	17	8.42
		Indian	0	0.00
		Kadazan-Dusun	2	0.99
		Others	0	0.00
β-Thalassaemia Intermedia	22	Malay	17	8.42
		Chinese	5	2.48
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
HbE/ β-Thalassaemia	76	Malay	69	34.16
		Chinese	3	1.49
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	4	1.98
HbH Disease	31	Malay	24	11.88
		Chinese	4	1.98
		Indian	1	0.50
		Kadazan-Dusun	0	0.00
		Others	2	0.99
Others	19	Malay	13	6.44
		Chinese	3	1.49
		Indian	1	0.50
		Kadazan-Dusun	0	0.00
		Others	2	0.99
Total			246	100.00

8.4 Treatment

8.4.1 Iron Chelation Therapy

Of the 202 patients in Negeri Sembilan, 135 patients (66.83%) receive iron chelation therapy. They are mainly β-thalassaemia major or HbE/β-thalassaemia patients. Most patients receive DFP (37 patients, 27.41%), an equal number of patients receive DFX or DFO (34 patients, 25.19%), and the remaining (30 patients, 22.22%) are on combination therapy (Table 8.7). Table 8.8 shows that most patients on DFP are receiving treatment at HTJ. Table 8.9 shows that most patients aged below 15 years old are on DFX, whereas patients aged 15 and above are on DFO, DFP or a combination of the two.

Table 8.7: Distribution of Patients in Negeri Sembilan by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	34	25.19
DFP only	37	27.41
DFX only	34	25.19
DFO + DFP	24	17.78
DFP + DFX	0	0.00
DFO + DFX	6	4.44
DFO + DFP + DFX	0	0.00
Total	135	100.00

Table 8.8: Distribution of Patients in Negeri Sembilan According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Jelebu	5	DFO only	4	2.96
		DFP only	1	0.74
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tuanku Ampuan Najihah, Kuala Pilah	17	DFO only	4	2.96
		DFP only	4	2.96
		DFX only	8	5.93
		DFO + DFP	1	0.74
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Port Dickson	3	DFO only	1	0.74
		DFP only	0	0.00
		DFX only	2	1.48
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tuanku Ja'afar	110	DFO only	25	18.52
		DFP only	32	23.70
		DFX only	24	17.78
		DFO + DFP	23	17.04
		DFP + DFX	0	0.00
		DFO + DFX	6	4.44
		DFO + DFP + DFX	0	0.00
Total			135	100.00

Table 8.9: Distribution of Patients in Negeri Sembilan According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	35	DFO only	5	14.29
		DFP only	1	2.86
		DFX only	27	77.14
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	2	5.71
		DFO + DFP + DFX	0	0.00
15 - 29.9	70	DFO only	23	32.86
		DFP only	17	24.29
		DFX only	5	7.14
		DFO + DFP	21	30.00
		DFP + DFX	0	0.00
		DFO + DFX	4	5.71
		DFO + DFP + DFX	0	0.00
30 - 44.9	20	DFO only	5	25.00
		DFP only	11	55.00
		DFX only	2	10.00
		DFO + DFP	2	10.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 44.9	8	DFO only	0	0.00
		DFP only	8	100.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	2	DFO only	1	50.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	50.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			135	

8.4.2 Serum Ferritin Level

Table 8.10 shows that 103 out of 137 transfused patients (75.18%) in Negeri Sembilan have their serum ferritin measured. The highest ferritin level recorded is 15,680.55 ng/mL (8-year-old patient diagnosed with β-thalassaemia major from Kuala Pilah). The percentage of patients with serum ferritin level lower than 2499 ng/mL is 46.60%, whereas the percentage of patients with serum ferritin above 2500 ng/mL is 53.40%.

Of the 137 regularly transfused patients, 22 of them are lost to follow-up, 2 had their last serum ferritin level taken in December 2018, and 10 had undergone bone marrow transplant, therefore no serum ferritin level was taken.

Table 8.10: Distribution of Patients in Negeri Sembilan According Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Jelebu	4	1	0.97	1	0.97	0	0.00	1	0.97	1	0.97
Hospital Tuanku Ampuan Najihah, Kuala Pilah	13	0	0.00	3	2.91	5	4.85	4	3.88	1	0.97
Hospital Port Dickson	3	0	0.00	2	1.94	1	0.97	0	0.00	0	0.00
Hospital Tuanku Ja'afar	83	14	13.59	27	26.21	25	24.27	15	14.56	2	1.94
Total	103	15	14.56	33	32.04	31	30.10	20	19.42	4	3.88

8.5 Conclusion

HTJ, Seremban and HTAN, Kuala Pilah serve as referral centres for thalassaemia patients in Negeri Sembilan. Patients receiving treatment from district hospitals are under the care of visiting specialists.

The age group with the highest number of thalassaemia patients in Negeri Sembilan is 15-29.9 years old. The patients' age range between 2 to 66 years old. Sixty-nine patients (34.18%) are below 18 years of age and is under paediatric care. Majority of the patients in Negeri Sembilan are female, with a male to female ratio of 1:1.5. Malay patients constitute 78.22% of the total number of patients, followed by Chinese (15.84%), Orang Asli (2.97%), Indian (0.99%), Kadazan-Dusun (0.99%) and other ethnicities (1%).

TDT patients in Negeri Sembilan are typically diagnosed with HbE/β-thalassaemia (37.62%) followed by β-thalassaemia major (26.73%). On the other hand, NTDT patients are diagnosed with HbH disease (15.35%), followed by β-thalassaemia intermedia (10.89%). Other diagnoses, such as other types of α-thalassaemia and HbS disease form 9.41% of the diagnoses. The oldest surviving β-thalassaemia major patient is above 60 years old.

Only 66.83% of the 202 patients in Negeri Sembilan are receiving iron chelation therapy, out of which 25.93% are under paediatric care. The remaining 33.17% of patients in Negeri Sembilan are not receiving any form of chelator.

Finally, despite chelation therapy and monitoring, only 75.18% of regularly transfused patients have their serum ferritin level measured. Of these, only 14.56% have a serum ferritin level below 1000 ng/mL, 32.04% have a serum ferritin level between 1000-2499 ng/mL, and 3.88% have a serum ferritin level more than 10,000 ng/mL.

9 Pahang

9.1 Introduction

The state of Pahang has a total of 11 hospitals; consisting of a state hospital (Hospital Tengku Ampuan Afzan [HTAA], Kuantan), a major hospital with specialist (Hospital Sultan Haji Ahmad Shah [HOSHAS], Temerloh), 2 minor hospitals with specialists (Hospital Kuala Lipis and Hospital Pekan) and 7 district hospitals.

Only 9 hospitals are included in the registry, namely HTAA, HOSHAS, Hospital Raub, Hospital Kuala Lipis, Hospital Jerantut, Hospital Jengka, Hospital Bentong, Hospital Pekan and Hospital Rompin. Cases from Hospital Muadzam Shah are referred to Hospital Rompin, whereas cases from Hospital Cameron Highlands are referred to Hospital Ipoh. Currently, there are no thalassaemia patients from Hospital Jengka.

The total population of Pahang is 1.67 million (Department of Statistics, Malaysia, 2019). There is a total of 432 thalassaemia patients in the state of Pahang.

Transfusion facilities have markedly improved in HTAA and HOSHAS, where all patients receive blood transfusions in day care and receive filtered and nucleic acid test (NAT)-tested blood. Other district hospitals still require ward admissions for transfusion and also provide filtered blood.

With the recent advances in thalassaemia service, HTAA is now able to provide MRI T2* facilities since October 2017. Bone densitometry examination is also provided under the Radiology Department in HTAA.

9.2 Patient Demographics

Data analysis includes patients who are either living, lost to follow-up or cured by transplant, and excludes deceased patients. In 2019, the total number of living patients in Pahang is 432 patients, and 8 deaths were reported. There are 217 regularly transfused patients (50.23%) and 215 irregularly transfused patients (49.77%).

Table 9.1: Distribution of Patients in Pahang by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Tengku Ampuan Afzan	259	59.95
Hospital Sultan Haji Ahmad Shah	119	27.55
Hospital Kuala Lipis	17	3.94
Hospital Pekan	13	3.01
Hospital Raub	5	1.16
Hospital Bentong	9	2.08
Hospital Jerantut	6	1.39
Hospital Jengka	0	0.00
Hospital Muadzam Shah	4	0.93
Hospital Sultanah Hajjah Kalsom	0	0.00
Total	432	100.00

Majority of thalassaemia patients in Pahang receive treatment at tertiary centres, i.e. HTAA (259 patients, 59.95%) and HOSHAS (119 patients, 27.55%).

Table 9.2: Distribution of Patients in Pahang by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	377
Cured by Stem Cell Therapy	2
Total	379
Lost to Follow-up	53
Total	432
Deaths in 2019	8
Cumulative Reported Deaths	48

Two of the 8 deaths in 2019 were due to cardiac causes. Four other deaths were due to infection, specifically severe sepsis. One 58-year-old female succumbed to renal complication, and a 41-year-old female died at home without medical post mortem (exact cause of death unknown).

Table 9.3: Cumulative Causes of Death in Pahang Since 2007

Causes of Death	Number of Patients	Percentage (%)
Cardiac Causes	7	15.91
Infections	26	59.09
Arrhythmias	1	2.27
Liver Disease	2	4.55
Motor Vehicle Accident (MVA)	1	2.27
Renal Complication	1	2.27
Tumours	3	6.82
Surgical Complication	1	2.27
Others	2	4.55
Total	44	100.00

Two patients classified under Other Causes of Death passed away from pulmonary haemorrhage and hypovolemic shock secondary to intra-abdominal bleeding. Since 2007, 4 patients had unknown causes of death (case notes were already disposed of by the hospital record department or died at home without post-mortem).

Table 9.4: New Thalassaemia Cases and New Thalassaemia Births by Year in Pahang

Year	New Cases	New Births
2014	37	11
2015	44	9
2016	43	4
2017	25	2
2018	17	1
2019	3	0

9.2.1 Age Group

Figure 9.1 indicates that patients aged 30 years and below form 75.23% (325 out of 432 patients) of the thalassaemia cases in Pahang. The eldest patient in Pahang is an 88-year-old diagnosed with HbH disease.

Figure 9.1: Distribution of Patients in Pahang by Age Group

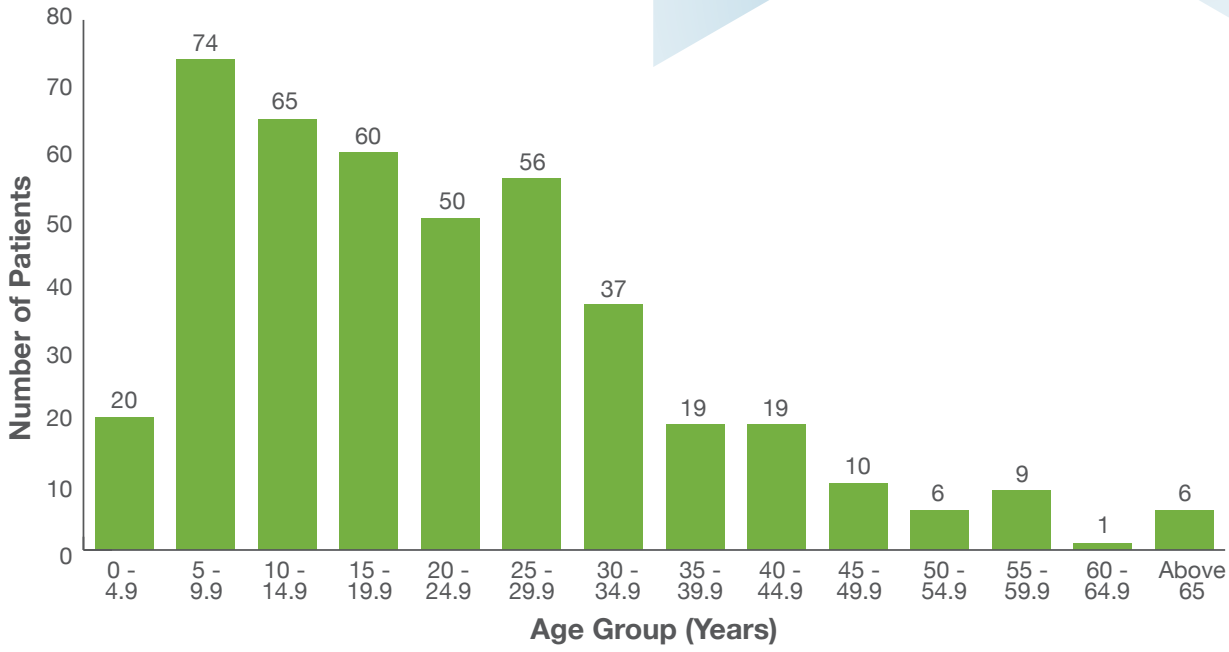


Table 9.5: Distribution of Patients in Pahang According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	159	β-Thalassaemia Major	15	9.43
		β-Thalassaemia Intermedia	8	5.03
		HbE/β-Thalassaemia	67	42.14
		HbH Disease	49	30.82
		Others	20	12.58
15 -29.9	166	β-Thalassaemia Major	37	22.29
		β-Thalassaemia Intermedia	12	7.23
		HbE/β-Thalassaemia	75	45.18
		HbH Disease	32	19.28
		Others	10	6.02
30 - 44.9	75	β-Thalassaemia Major	11	14.67
		β-Thalassaemia Intermedia	6	8.00
		HbE/β-Thalassaemia	35	46.67
		HbH Disease	15	20.00
		Others	8	10.67
45 - 59.9	25	β-Thalassaemia Major	3	12.00
		β-Thalassaemia Intermedia	5	20.00
		HbE/β-Thalassaemia	6	24.00
		HbH Disease	7	28.00
		Others	4	16.00

60 and above	7	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	2	28.57
		HbH Disease	4	57.14
		Others	1	14.29
Total			432	

9.2.2 Gender

Table 9.6 shows that female patients dominate the population of thalassaemia patients in Pahang, comprising 57.21% of the total number of patients (250 patients), whereas male patients form the remaining 41.65% (182 patients).

Table 9.6: Distribution of Patients in Pahang According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Tengku Ampuan Afzan	100	23.15	159	36.81
Hospital Sultan Haji Ahmad Shah	58	13.43	61	14.12
Hospital Kuala Lipis	10	2.31	7	1.62
Hospital Pekan	5	1.16	8	1.85
Hospital Raub	1	0.23	4	0.93
Hospital Bentong	4	0.93	5	1.16
Hospital Jerantut	2	0.46	4	0.93
Hospital Jengka	0	0.00	0	0.00
Hospital Muadzam Shah	2	0.46	2	0.46
Hospital Sultanah Hajjah Kalsom	0	0.00	0	0.00
Total	182	42.13	250	57.87

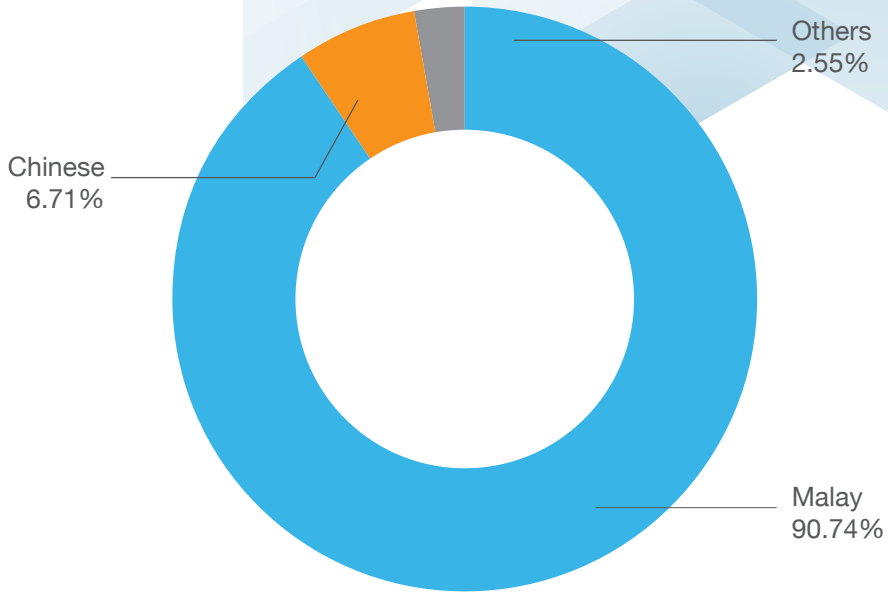
9.2.3 Ethnic Group

Based on Figure 9.2, a majority of thalassaemia patients in Pahang are of Malay descent (392 patients, 90.74%), followed by Chinese (29 patients, 6.71%) and other ethnicities (11 patients, 2.55%). Table 9.9 summarises the distribution of patient ethnicities according to centre.

Table 9.7: Distribution of Patients in Pahang According to Ethnic Group by Centre

Centre	Total	Malay		Chinese		Indian		Others	
		No.	%	No.	%	No.	%	No.	%
Hospital Tengku Ampuan Afzan	259	239	55.32	18	4.17	0	0.00	2	0.46
Hospital Sultan Haji Ahmad Shah	119	108	25.00	5	1.16	0	0.00	6	1.39
Hospital Kuala Lipis	17	16	3.70	0	0.00	0	0.00	1	0.23
Hospital Pekan	13	12	2.78	0	0.00	0	0.00	1	0.23
Hospital Raub	5	4	0.93	1	0.23	0	0.00	0	0.00
Hospital Bentong	9	5	1.16	3	0.69	0	0.00	1	0.23
Hospital Jerantut	6	5	1.16	1	0.23	0	0.00	0	0.00
Hospital Jengka	0	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Muadzam Shah	4	3	0.69	1	0.23	0	0.00	0	0.00
Total	432	392	90.74	29	6.71	0	0.00	11	2.55

Figure 9.2: Distribution of Patients in Pahang by Ethnic Group



9.3 Diagnosis

As shown in Figure 9.3, the diagnosis with the highest number of patients in Pahang is HbE/β-thalassaemia (185 patients, 42.82%), followed by HbH disease (107 patients, 24.77%), β-thalassaemia major (66 patients, 15.28%), other diagnoses (43 patients, 9.95%) and β-thalassaemia intermedia (31 patients, 7.18%).

Figure 9.3: Distribution of Patients in Pahang by Diagnosis

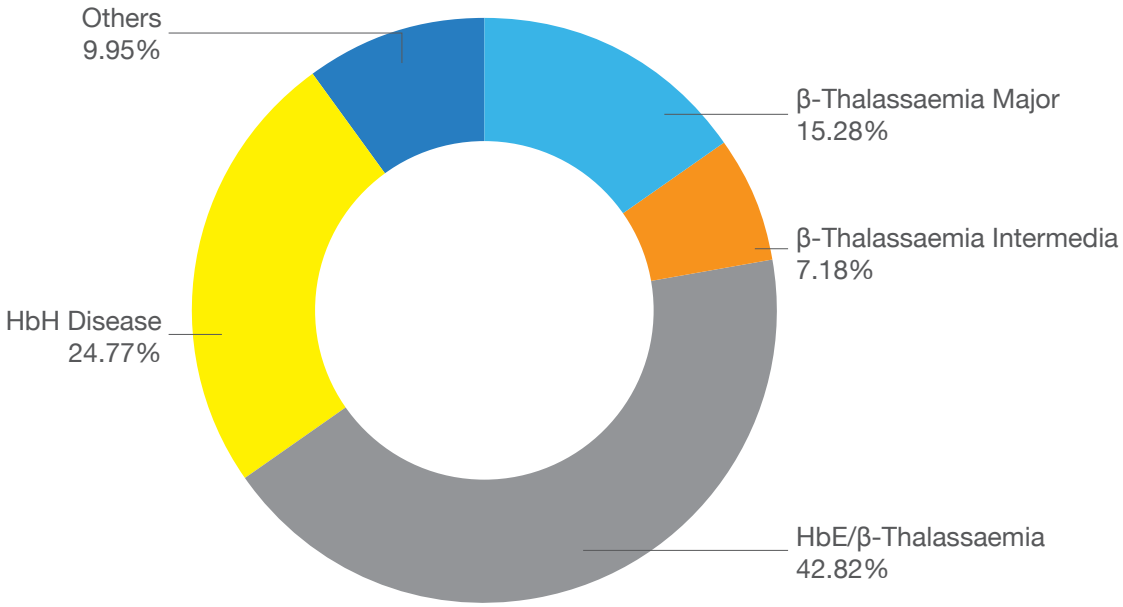


Table 9.8: Distribution of Patients in Pahang According to Diagnosis by Centre

Centre	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/ β-Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Tengku Ampuan Afzan	29	6.71	19	4.40	119	27.55	64	14.81	28	6.48
Hospital Sultan Haji Ahmad Shah	19	4.40	11	2.55	53	12.27	29	6.71	7	1.62
Hospital Kuala Lipis	9	2.08	1	0.23	4	0.93	1	0.23	2	0.46
Hospital Pekan	0	0.00	0	0.00	2	0.46	7	1.62	4	0.93
Hospital Raub	2	0.46	0	0.00	1	0.23	1	0.23	1	0.23
Hospital Bentong	2	0.46	0	0.00	3	0.69	4	0.93	0	0.00
Hospital Jerantut	4	0.93	0	0.00	1	0.23	1	0.23	0	0.00
Hospital Jengka	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Muadzam Shah	1	0.23	0	0.00	2	0.46	0	0.00	1	0.23
Hospital Sultanah Hajjah Kalsom	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	**66**	**15.28**	**31**	**7.18**	**185**	**42.82**	**107**	**24.77**	**43**	**9.95**

Table 9.9: Distribution of Patients in Pahang According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	66	Malay	58	13.43
		Chinese	8	1.85
		Indian	0	0.00
		Others	0	0.00
β-Thalassaemia Intermedia	31	Malay	30	6.94
		Chinese	1	0.23
		Indian	0	0.00
		Others	0	0.00
HbE/ β-Thalassaemia	185	Malay	172	39.81
		Chinese	9	2.08
		Indian	0	0.00
		Others	4	0.93
HbH Disease	107	Malay	96	22.22
		Chinese	8	1.85
		Indian	0	0.00
		Others	3	0.69
Others	43	Malay	36	8.33
		Chinese	3	0.69
		Indian	0	0.00
		Others	4	0.93
Total			432	100.00

9.4 Treatment

9.4.1 Iron Chelation Therapy

Based on Table 9.10, a total of 253 patients in Pahang receive iron chelating agents. Twenty-eight patients (11.07%) receive DFO, 89 patients (35.18%) receive DFP, 66 patients (26.09%) receive DFX, 53 patients (20.95%) receive a combination of DFO and DFP, 3 patients (1.19 %) receive a combination of DFP and DFX, and 13 patients (5.14%) receive a combination of DFO and DFX. One patient receive a combination of all three iron chelators (DFO, DFP and DFX).

Table 9.10: Distribution of Patients in Pahang by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	28	11.07
DFP only	89	35.18
DFX only	66	26.09
DFO + DFP	53	20.95
DFP + DFX	3	1.19
DFO + DFX	13	5.14
DFO + DFP + DFX	1	0.40
Total	253	100.00

Table 9.11: Distribution of Patients in Pahang According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Tengku Ampuan Afzan, Kuantan	147	DFO only	16	6.32
		DFP only	52	20.55
		DFX only	34	13.44
		DFO + DFP	33	13.04
		DFP + DFX	2	0.79
		DFO + DFX	9	3.56
		DFO + DFP + DFX	1	0.40
Hospital Sultan Haji Ahmad Shah	83	DFO only	11	4.35
		DFP only	29	11.46
		DFX only	22	8.70
		DFO + DFP	17	6.72
		DFP + DFX	1	0.40
		DFO + DFX	3	1.19
		DFO + DFP + DFX	0	0.00
Hospital Kuala Lipis	12	DFO only	1	0.40
		DFP only	5	1.98
		DFX only	6	2.37
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Pekan	1	DFO only	0	0.00
		DFP only	1	0.40
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Raub	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	0.40
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Bentong	5	DFO only	0	0.00
		DFP only	1	0.40
		DFX only	3	1.19
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.40
		DFO + DFP + DFX	0	0.00
Hospital Jerantut	4	DFO only	0	0.00
		DFP only	1	0.40
		DFX only	0	0.00
		DFO + DFP	3	1.19
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			253	100.00

Table 9.12: Distribution of Patients in Pahang According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0–14.9	53	DFO only	2	3.77
		DFP only	6	11.32
		DFX only	43	81.13
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	2	3.77
		DFO + DFP + DFX	0	0.00
15–29.9	122	DFO only	17	13.93
		DFP only	40	32.79
		DFX only	21	17.21
		DFO + DFP	33	27.05
		DFP + DFX	1	0.82
		DFO + DFX	9	7.38
		DFO + DFP + DFX	1	0.82

30–44.9	56	DFO only	7	12.50
		DFP only	26	46.43
		DFX only	2	3.57
		DFO + DFP	17	30.36
		DFP + DFX	2	3.57
		DFO + DFX	2	3.57
		DFO + DFP + DFX	0	0.00
45–59.9	18	DFO only	2	11.11
		DFP only	13	72.22
		DFX only	0	0.00
		DFO + DFP	3	16.67
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	4	DFO only	0	0.00
		DFP only	4	100.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			253	

9.4.2 Serum Ferritin Level

There are 149 patients in Pahang who have their serum ferritin level measured in 2019. Table 9.13 shows the distribution of patients according to their most recent serum ferritin level in each centre in Pahang. One hundred and four patients (69.79%) have a serum ferritin level below 2499 ng/mL and 45 patients (30.20%) have a serum ferritin level above 2500 ng/mL. Only 41 patients (27.52%) achieve a serum ferritin level below 1000 ng/mL.

Table 9.13: Distribution of Patients in Pahang According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No	%	No	%	No	%	No	%	No	%
Hospital Tengku Ampuan Afzan	96	29	19.46	30	20.13	25	16.78	12	8.05	0	0.00
Hospital Sultan Haji Ahmad Shah	52	11	7.38	33	22.15	5	3.36	2	1.34	1	0.67
Hospital Kuala Lipis	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Pekan	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Raub	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Bentong	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Jerantut	1	1	0.67	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Jengka	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Muadzam Shah	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	149	41	27.52	63	42.28	30	20.13	14	9.40	1	0.67

9.5 Conclusion

There are 182 (41.65%) male patients and 250 (57.21%) female patients in Pahang. Patients aged between 6-10 years old form the largest age group of patients in Pahang. The number of thalassaemia patients increases from age groups above 6 years old and reduces in older age groups.

There are 66 (15.28%) patients diagnosed with β-thalassaemia major in Pahang. Malay patients form a majority of the patients in Pahang (392 patients, 90.74%), followed by Chinese patients (29 patients, 6.71%) and patients of other ethnicities (11 patients, 2.55%).

A total of 253 patients in Pahang receive iron chelating agents. One patient receive a combination of all 3 iron chelators (DFO, DFP and DFX). There are 149 patients in Pahang who have their serum ferritin levels measured in 2019. Of these, 45 patients (30.20%) have a serum ferritin level above 2500 ng/mL.

10 Perak

10.1 Introduction

Perak is 1 of the 13 states in Malaysia, the fourth largest state in the country and the second largest state in Peninsular Malaysia. It has an estimated population of 2.548 million people (Department of Statistics Malaysia, 2017). Perak is divided into 10 administrative districts, namely Hulu Perak, Larut & Matang, Kerian, Kuala Kangsar, Kinta, Kampar, Perak Tengah, Manjung, Hilir Perak, and Batang Padang. Hospital facilities are provided in each district, so patients may receive treatment conveniently. However, thalassaemia patients are treated in only 7 hospitals, which are Hospital Raja Permaisuri Bainun (Ipoh), Hospital Taiping, Hospital Teluk Intan, Hospital Manjung, Hospital Kuala Kangsar, Hospital Slim River and Hospital Gerik.

10.2 Patient Demographics

There are 594 thalassaemia patients in Perak. These patients receive transfusions and care in the 7 hospitals listed in Table 10.1. Most of these hospitals carry out transfusions in their Ambulatory Care Centre (ACC); for hospitals without ACC, the transfusions are carried out in the wards. Table 10.1 also shows that most thalassaemia patients (348 patients, 58.59%) receive treatment at Hospital Raja Permaisuri Bainun.

Table 10.1: Distribution of Patients in Perak by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Raja Permaisuri Bainun	348	58.59
Hospital Taiping	104	17.51
Hospital Teluk Intan	56	9.43
Hospital Kuala Kangsar	15	2.53
Hospital Slim River	14	2.36
Hospital Gerik	13	2.19
Hospital Seri Manjung	44	7.41
Total	594	100.00

Table 10.2: Distribution of Patients in Perak by Vital Status

Vital Status	Number of Patients
Alive and on Active Treatment	540
Cured by Stem Cell Therapy (2008-2017)	8
Total	548
Lost to Follow-up	46
Total	594
Deaths in 2019	4
Cumulative Reported Deaths	47

Table 10.3: Cumulative Causes of Death in Perak Since 2007*

Causes of Death	Number of Patients	Percentage (%)
Cardiac Causes	23	56.10
Endocrine Complications	2	4.88
Tumours	1	2.44
Arrhythmias	1	2.44
Infections	10	24.39
Others	4	9.76
Total	41	100.00

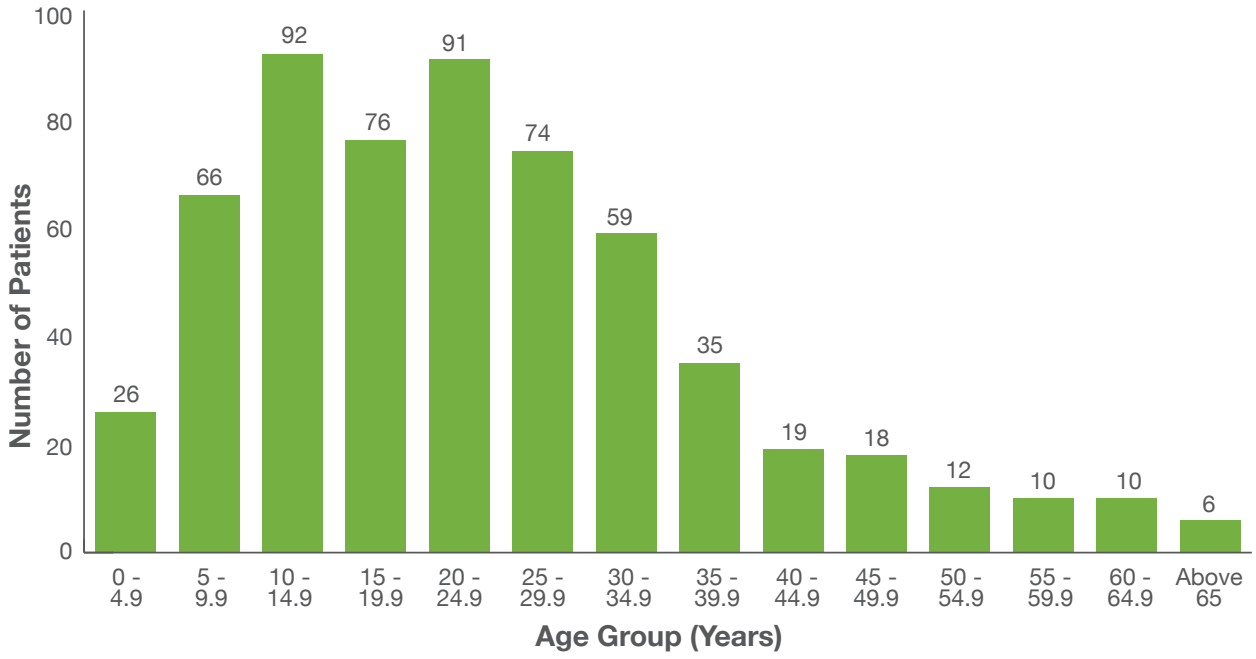
**Six patients had unavailable data and unknown causes of death.*

Four patients passed away in 2019. A 35-year-old female patient died due to cardiac causes, while another 3 patients died at home (their causes of death are unknown).

10.2.1 Age Group

Based on Figure 10.1, 43.26% of the thalassaemia patients in Perak are 20 years old and below, and are in the paediatric and adolescent age groups. Patients above 20 years old account for 56.74% of the thalassaemics in Perak.

Figure 10.1: Distribution of Patients in Perak by Age Group



As seen in Figure 10.1, the majority of thalassaemia patients in Perak are within the second and third decades of life.

Table 10.4: Distribution of Patients in Perak According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	184	β-Thalassaemia Major	41	22.28
		β-Thalassaemia Intermedia	14	7.61
		HbE/β-Thalassaemia	87	47.28
		HbH Disease	34	18.48
		Others	8	4.35
15 - 29.9	241	β-Thalassaemia Major	57	23.65
		β-Thalassaemia Intermedia	24	9.96
		HbE/β-Thalassaemia	107	44.40
		HbH Disease	49	20.33
		Others	4	1.66
30 - 44.9	113	β-Thalassaemia Major	25	22.12
		β-Thalassaemia Intermedia	9	7.96
		HbE/β-Thalassaemia	43	38.05
		HbH Disease	26	23.01
		Others	10	8.85
45 - 59.9	40	β-Thalassaemia Major	1	2.50
		β-Thalassaemia Intermedia	3	7.50
		HbE/β-Thalassaemia	18	45.00
		HbH Disease	16	40.00
		Others	2	5.00
60 and above	16	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	3	18.75
		HbE/β-Thalassaemia	4	25.00
		HbH Disease	7	43.75
		Others	2	12.50
Total			594	

10.2.2 Gender

Table 10.5 shows the distribution of thalassaemia patients in Perak by gender. There are 311 (52.36%) and 283 (47.64%) male and female patients, respectively.

Table 10.5: Distribution of Patients in Perak According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Raja Permaisuri Bainun	165	27.78	183	30.81
Hospital Taiping	59	9.93	45	7.58
Hospital Teluk Intan	37	6.23	19	3.20
Hospital Kuala Kangsar	8	1.35	7	1.18
Hospital Slim River	9	1.52	5	0.84
Hospital Gerik	6	1.01	7	1.18
Hospital Seri Manjung	27	4.55	17	2.86
Total	311	52.36	283	47.64

10.2.3 Ethnic Group

Patients of Malay descent comprise the largest cohort of thalassaemics in Perak, with 458 patients (77.10%), followed by the Chinese (112 patients, 18.86%). Other ethnic groups contributed to only a minor proportion of the total number of thalassaemics in Perak; this includes Indian (3 patients, 0.51%), Kadazan (1 patient, 0.18%), Orang Asli (17 patients, 2.86%) and other ethnic groups (4 patients, 0.67%).

Figure 10.2: Distribution of Patients in Perak by Ethnic Group

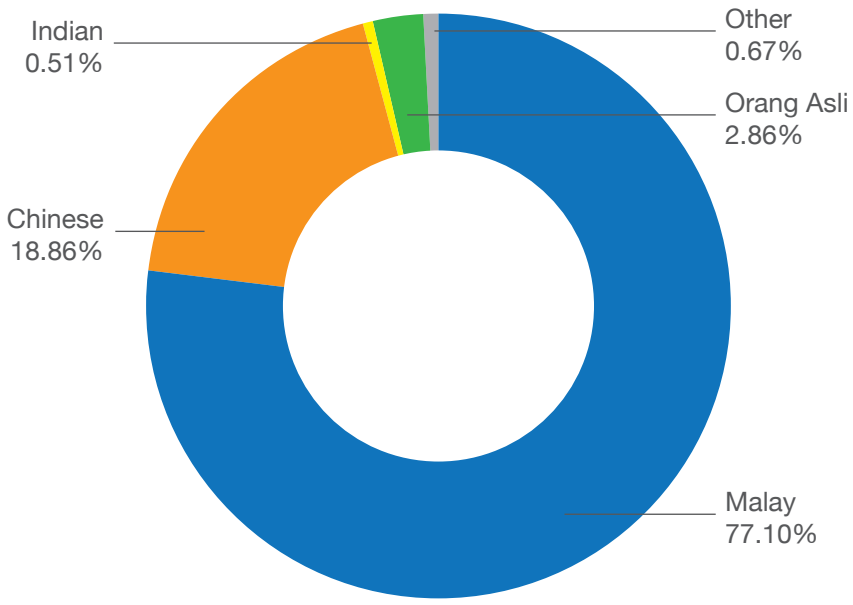


Table 10.6: Distribution of Patients in Perak According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Orang Asli		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Raja Permaisuri Bainun	239	40.24	96	16.16	2	0.34	10	1.68	1	0.17
Hospital Taiping	98	16.50	6	1.01	0	0.00	0	0.00	0	0.00
Hospital Teluk Intan	44	7.41	6	1.01	1	0.17	4	0.67	1	0.17
Hospital Kuala Kangsar	15	2.53	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Slim River	12	2.02	0	0.00	0	0.00	0	0.00	2	0.34
Hospital Gerik	10	1.68	0	0.00	0	0.00	3	0.51	0	0.00
Hospital Seri Manjung	40	6.73	4	0.67	0	0.00	0	0.00	0	0.00
Total	458	77.10	112	18.86	3	0.51	17	2.86	4	0.67

10.3 Diagnosis

HbE/β-thalassaemia is the main diagnosis in Perak with 259 patients (43.60%), followed by HbH disease (132 patients, 22.22%), β-thalassaemia major (124 patients, 20.88%) and β-thalassaemia intermedia (53 patients, 8.92%). The remaining 26 patients (4.38%) have other forms of haemoglobinopathy such as α-thalassaemia, Hb Adana, δβ variant and homozygous HbE.

Figure 10.3: Distribution of Patients in Perak by Diagnosis

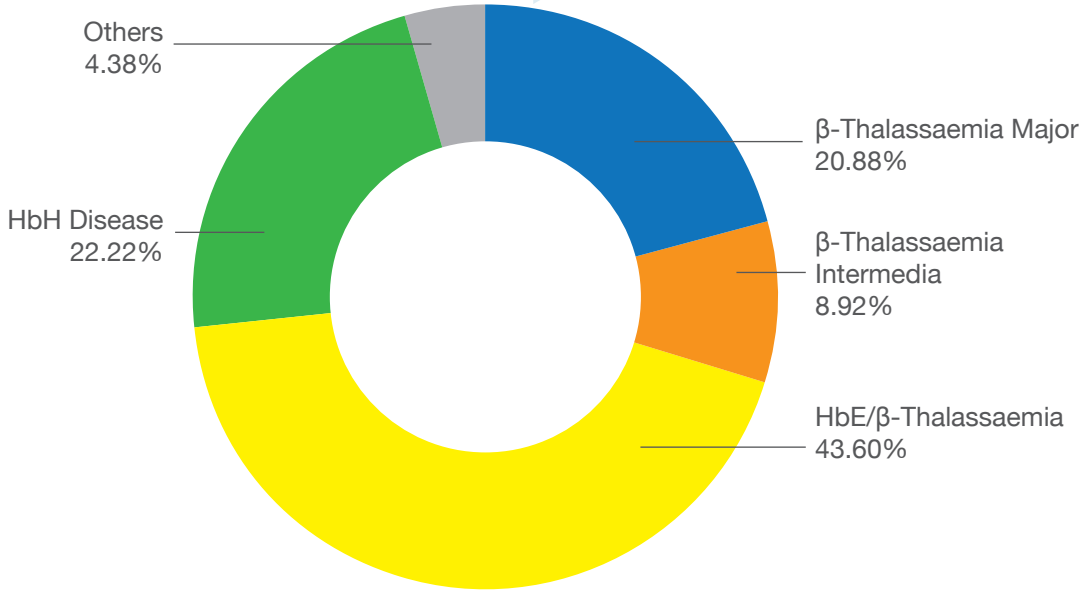


Table 10.7: Distribution of Patients in Perak According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	124	Malay	86	14.48
		Chinese	35	5.89
		Indian	1	0.17
		Orang Asli	1	0.17
		Others	1	0.17
β-Thalassaemia Intermedia	53	Malay	39	6.57
		Chinese	12	2.02
		Indian	1	0.17
		Orang Asli	1	0.17
		Others	0	0.00
HbE/ β-Thalassaemia	259	Malay	229	38.55
		Chinese	19	3.20
		Indian	0	0.00
		Orang Asli	8	1.35
		Others	3	0.51
HbH Disease	128	Malay	84	14.14
		Chinese	42	7.07
		Indian	0	0.00
		Orang Asli	6	1.01
		Others	0	0.00

Others*	30	Malay	20	3.37
		Chinese	4	0.67
		Indian	1	0.17
		Orang Asli	1	0.17
		Others	0	0.00
Total			594	100.00

10.4 Treatment

10.4.1 Iron Chelation Therapy

Based on Table 10.8, 329 patients in Perak received iron chelation therapy. Eighty-one patients (22.88%) are receiving DFO therapy only, and 72 patients (20.34%) are receiving oral DFP therapy only. Another 128 patients (36.16%) are on DFX, which is normally reserved for patients who could not tolerate other chelating agents. Furthermore, 55 patients (15.54%) are receiving a combination of DFO and DFP. In addition, the number of patients on a combination of DFP + DFX and DFO + DFX are 8 (2.26%) each. Only 2 patients (0.56%) are receiving a DFO + DFP + DFX combination. Nevertheless, a high proportion of patients in Perak (240 patients, 40.40%) are not on iron chelation therapy.

Table 10.8: Distribution of Patients in Perak by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	81	22.88
DFP only	72	20.34
DFX only	128	36.16
DFO + DFP	55	15.54
DFP + DFX	8	2.26
DFO + DFX	8	2.26
DFO + DFP + DFX	2	0.56
Total	354	100.00

Table 10.9: Distribution of Patients in Perak According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Raja Permaisuri Bainun	180	DFO only	44	12.43
		DFP only	47	13.28
		DFX only	54	15.25
		DFO + DFP	29	8.19
		DFP + DFX	4	1.13
		DFO + DFX	0	0.00
		DFO + DFP + DFX	2	0.56
Hospital Taiping	75	DFO only	9	2.54
		DFP only	15	4.24
		DFX only	28	7.91
		DFO + DFP	16	4.52
		DFP + DFX	3	0.85
		DFO + DFX	4	1.13
		DFO + DFP + DFX	0	0.00

Hospital Teluk Intan	39	DFO only	10	2.82
		DFP only	5	1.41
		DFX only	17	4.80
		DFO + DFP	6	1.69
		DFP + DFX	0	0.00
		DFO + DFX	1	0.28
		DFO + DFP + DFX	0	0.00
Hospital Kuala Kangsar	11	DFO only	4	1.13
		DFP only	2	0.56
		DFX only	4	1.13
		DFO + DFP	1	0.28
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Slim River	11	DFO only	3	0.85
		DFP only	0	0.00
		DFX only	8	2.26
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Gerik	5	DFO only	2	0.56
		DFP only	0	0.00
		DFX only	3	0.85
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Seri Manjung	33	DFO only	9	2.54
		DFP only	3	0.85
		DFX only	14	3.95
		DFO + DFP	3	0.85
		DFP + DFX	1	0.28
		DFO + DFX	3	0.85
		DFO + DFP + DFX	0	0.00
Total			354	100.00

Table 10.10: Distribution of Patients in Perak According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	96	DFO only	15	15.63
		DFP only	0	0.00
		DFX only	79	82.29
		DFO + DFP	1	1.04
		DFP + DFX	0	0.00
		DFO + DFX	1	1.04
		DFO + DFP + DFX	0	0.00
15 - 29.9	153	DFO only	43	28.10
		DFP only	22	14.38
		DFX only	39	25.49
		DFO + DFP	35	22.88
		DFP + DFX	6	3.92
		DFO + DFX	6	3.92
		DFO + DFP + DFX	2	1.31
30 - 44.9	75	DFO only	18	24.00
		DFP only	33	44.00
		DFX only	8	10.67
		DFO + DFP	15	20.00
		DFP + DFX	1	1.33
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 44.9	17	DFO only	3	17.65
		DFP only	10	58.82
		DFX only	2	11.76
		DFO + DFP	2	11.76
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	13	DFO only	2	15.38
		DFP only	7	53.85
		DFX only	0	0.00
		DFO + DFP	2	15.38
		DFP + DFX	1	7.69
		DFO + DFX	1	7.69
		DFO + DFP + DFX	0	0.00
Total			354	

10.4.2 Serum Ferritin Level

A total 243 TDT patients (64.63%) in Perak had their most recent ferritin level measured between January and October 2019. About 50 patients (20.58%) have a serum ferritin level at an ideal level which is below 1000 ng/mL. Eighty-three patients (34.16%) have a serum ferritin level between 1000 to 2499 ng/mL, whereas 110 patients (45.27%) have a serum ferritin level above 2500 ng/mL.

Table 10.11: Distribution of Patients in Perak According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No	%	No	%	No	%	No	%	No	%
Hospital Raja Permaisuri Bainun	126	27	11.11	43	17.70	44	18.11	11	4.53	1	0.41
Hospital Taiping	56	8	3.29	19	7.82	13	5.35	10	4.12	6	2.47
Hospital Teluk Intan	25	5	2.06	10	4.12	8	3.29	2	0.82	0	0.00
Hospital Kuala Kangsar	2	0	0.00	1	0.41	1	0.41	0	0.00	0	0.00
Hospital Slim River	10	5	2.06	3	1.23	2	0.82	0	0.00	0	0.00
Hospital Gerik	4	1	0.41	0	0.00	2	0.82	1	0.41	0	0.00
Hospital Seri Manjung	20	4	1.65	7	2.88	5	2.06	3	1.23	1	0.41
Total	243	50	20.58	83	34.16	75	30.86	27	11.11	8	3.29

10.5 Conclusion

This chapter provides a brief overview on the number of thalassaemia patients, whom received treatment in the state of Perak. In 2019, the number of thalassaemia patients reported was 594, of which TDT and NTDT comprise 298 (50.17%) and 296 (49.83%) patients, respectively.

The majority of thalassaemia patients in Perak are adults (373 patients, ,62.79%) followed by children (221 patients, 37.21%). Malay patients constitute 74.65% of the total number of patients, followed by Chinese (20.04%), and other ethnicities (5.32%). The subtypes of thalassaemia found in the state are mainly HbE/β-thalassaemia (43.60%), followed by HbH disease (22.22%), β-thalassaemia major (20.88%) and β-thalassaemia intermedia (8.92%).

The total number of patients on iron chelation therapy is 354 patients (59.60%), which includes TDT and NTDT patients. A total of 266 TDT patients (89.26%) are currently receiving chelation therapy. The remaining 10.74% of TDT patients are not on chelation therapy, mainly due to non-compliance to treatment. A total of 88 NTDT patients (29.73%) are also receiving chelation therapy. The main type of chelator used is DFX (36.16%), followed by DFO (22.88%). This is mainly due to its use in the paediatric age group, which favours oral iron chelators to subcutaneous preparation. The most common combination therapy is DFO + DFP (15.54%).

Serum ferritin levels below 1000 ng/mL indicate good compliance to iron chelators; however, only 20.58% of TDT patients fall into this category. A total of 243 (81.54%) TDT patients had their serum ferritin levels measured, of which 54.74% have an acceptable serum ferritin level of lower than 2500 ng/mL. Thus, compliance still remains an issue in 110 patients (45.27%) in Perak.

Several measures have been undertaken to improve these statistics, among which are to consolidate treatment in major hospitals with specialists or at least with regular visiting specialists, and to set up a dedicated team to manage thalassaemia patients.

11 Perlis

11.1 Introduction

Perlis is the smallest state in Malaysia, with an estimated population of 257,300 as of 2019 (Department of Statistics Malaysia). There is only one hospital in Perlis, which is Hospital Tuanku Fauziah (HTF) located in Kangar.

11.2 Patient Demographics

Data analysed were taken from patients who are either alive, lost to follow-up or cured by transplant; however, deceased patients are excluded. There are 126 living patients in Perlis, and 14 patients are deceased. Two patients passed away in 2019 due to infection and acute leukaemia, respectively.

Table 11.1: Distribution of Patients in Perlis by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Tunku Fauziah, Kangar	126	100.00
Total	126	100.00

Table 11.2: Distribution of Patients in Perlis by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	109
Cured by Stem Cell Therapy	1
Total	110
Lost to Follow-up	16
Total	126
Deaths in 2019	2
Cumulative Reported Deaths	14

Table 11.3: Cumulative Causes of Death in Perlis Since 2007

Causes of Death	Number of Patients	Percentage (%)
Infections	3	21.43
Cardiac Causes	7	50.00
Motor Vehicle Accident	3	21.43
Tumour	1	7.14
Total	14	100.00

11.2.1 Age Group

The youngest patient in Perlis is 2 years old and the eldest is 64 years old. Sixty-two patients (49.21%) are below 20 years old. The median age group is 20–24.9 years old.

Figure 11.1: Distribution of Patients in Perlis by Age Group

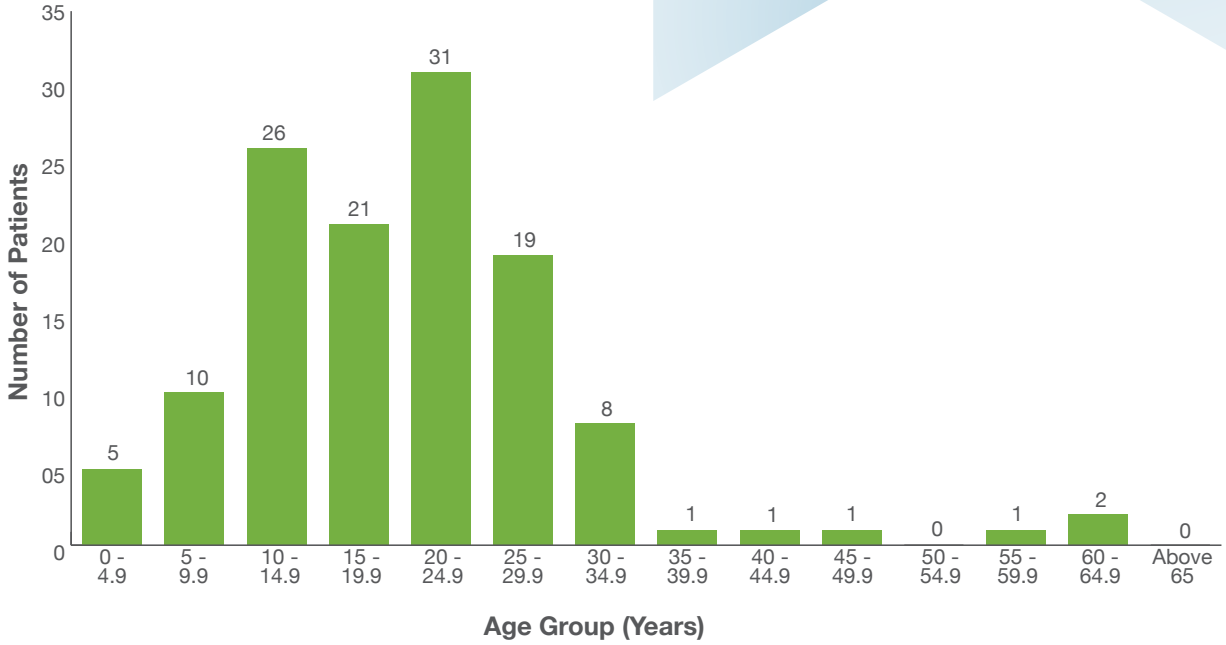


Table 11.4: Distribution of Patients in Perlis According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	41	β-Thalassaemia Major	10	24.39
		β-Thalassaemia Intermedia	10	24.39
		HbE/β-Thalassaemia	10	24.39
		HbH Disease	9	21.95
		Others	2	4.88
15 - 29.9	71	β-Thalassaemia Major	13	18.31
		β-Thalassaemia Intermedia	6	8.45
		HbE/β-Thalassaemia	36	50.70
		HbH Disease	15	21.13
		Others	1	1.41
30 - 44.9	10	β-Thalassaemia Major	4	40.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	6	60.00
		HbH Disease	0	0.00
		Others	0	0.00

45 - 59.9	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	50.00
		HbH Disease	0	0.00
		Others	1	50.00
60 and above	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	50.00
		HbH Disease	0	0.00
		Others	1	50.00
Total			126	

11.2.2 Gender

Table 11.5 shows the distribution of thalassaemia patients in Perlis by gender. The number of male patients is higher (67 patients, 53.17%) compared to female patients (59 patients, 46.83%).

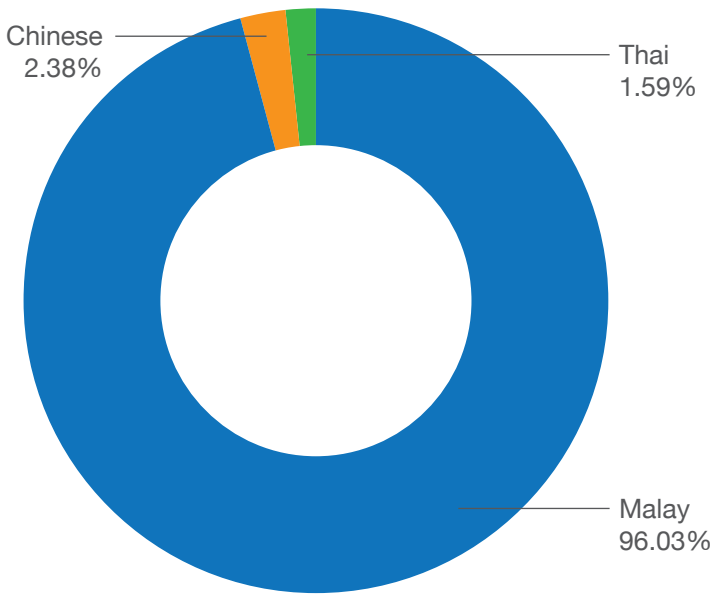
Table 11.5: Distribution of Patients in Perlis by Gender

Centre	Male		Female	
	Number	%	Number	%
Hospital Tunku Fauziah	67	53.17	59	46.83
Total	67	53.17	59	46.83

11.2.3 Ethnic Group

As shown in Figure 11.2, most thalassaemia patients in Perlis are of Malay descent (121 patients, 96.03%). Three patients (2.38%) are Chinese and 2 patients are Thai (1.59%).

Figure 11.2: Distribution of Patients in Perlis by Ethnic Group



11.3 Diagnosis

The most common diagnosis among patients in Perlis is HbE/β-thalassaemia (54 patients, 42.86%), followed by β-thalassaemia major (27 patients, 21.43%), HbH disease (24 patients, 19.05%) and β-thalassaemia intermedia (16 patients, 12.70%). The remaining 5 patients (3.97%) have other diagnoses (Figure 11.3).

Figure 11.3: Distribution of Patients in Perlis by Diagnosis

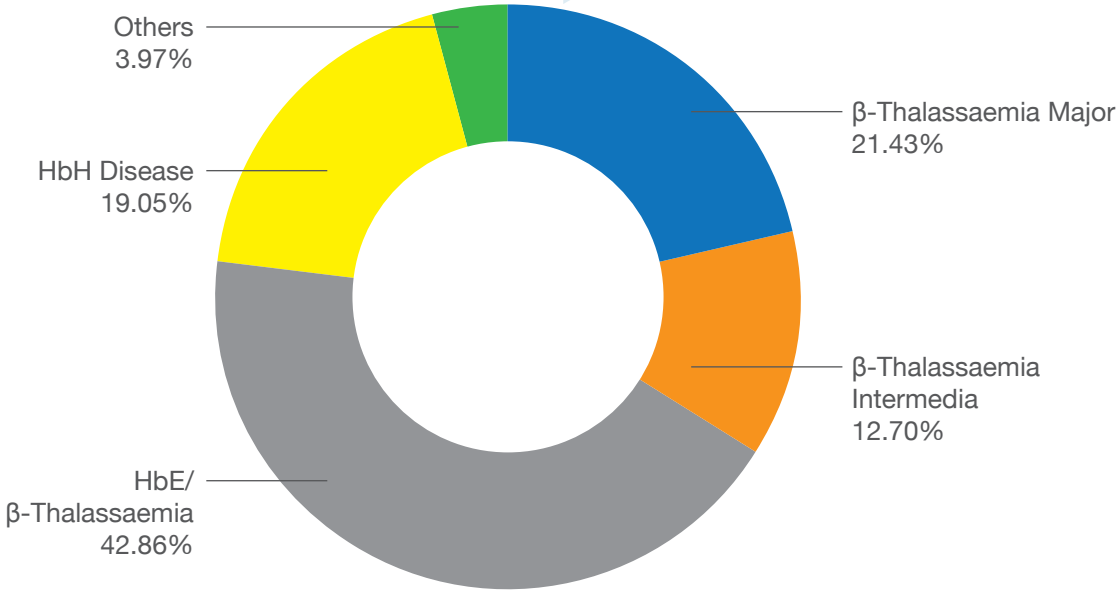


Table 11.6: Distribution of Patients in Perlis According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	27	Malay	26	20.63
		Chinese	1	0.79
		Indian	0	0.00
		Thai	0	0.00
β-Thalassaemia Intermedia	16	Malay	16	12.70
		Chinese	0	0.00
		Indian	0	0.00
		Thai	0	0.00
HbE/ β-Thalassaemia	54	Malay	52	41.27
		Chinese	1	0.79
		Indian	0	0.00
		Thai	1	0.79
HbH Disease	24	Malay	22	17.46
		Chinese	1	0.79
		Indian	0	0.00
		Thai	1	0.79
Others	5	Malay	5	3.97
		Chinese	0	0.00
		Indian	0	0.00
		Thai	0	0.00
Total			126	100.00

11.4 Treatment

11.4.1 Iron Chelation Therapy

Seventy-five patients (59.52%) in Perlis receive iron chelation therapy. Sixty-four (85.33%) of the chelated patients are on oral DFX, 3 patients (4%) are on subcutaneous DFO, 3 patients (4%) are on oral DFP and 5 patients (6.67%) are on combination therapy.

Table 11.7: Distribution of Patients in Perlis by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	3	4.00
DFP only	3	4.00
DFX only	64	85.33
DFO + DFP	2	2.67
DFP + DFX	1	1.33
DFO + DFX	2	2.67
DFO + DFP + DFX	0	0.00
Total	75	100.00

Table 11.8: Distribution of Patients in Perlis According to Type of Iron Chelator Received by Age Group

Age Group	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	18	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	17	94.44
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	5.56
		DFO + DFP + DFX	0	0.00
15 - 29.9	48	DFO only	3	6.25
		DFP only	1	2.08
		DFX only	41	85.42
		DFO + DFP	2	4.17
		DFP + DFX	1	2.08
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 44.9	7	DFO only	0	0.00
		DFP only	1	14.29
		DFX only	5	71.43
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	14.29
		DFO + DFP + DFX	0	0.00
45 - 59.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	100.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

60 and above	1	DFO only	0	0.00
		DFP only	1	100.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			75	

11.4.2 Serum Ferritin Level

There are 53 regularly transfused patients in Perlis who had their serum ferritin level measured. Thirty-two patients (60.38%) have serum ferritin levels lower than 2500 ng/mL, and 21 patients (39.62%) have moderate to severe iron overload, with serum ferritin levels above 2500 ng/mL.

Table 11.9: Distribution of Patients in Perlis According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No	%	No	%	No	%	No	%	No	%
Hospital Tunku Fauziah	53	10	18.87	22	41.51	12	22.64	8	15.09	1	1.89
Total	53	10	18.87	22	41.51	12	22.64	8	15.09	1	1.89

11.5 Conclusion

Approximately 49.21% of the thalassaemia patients in Perlis are below 20 years old. The median age group is 20-24.9 years old. Most patients (96.03%) are Malay, followed by Chinese (2.38%) and Thai (1.59%). The most common type of thalassaemia in Perlis is HbE/β-thalassaemia, followed by β-thalassaemia major and HbH disease. Sixty-four (85.33%) of the chelated patients are on oral DFX and most of the regularly transfused patients (60.38%) are well-chelated with a serum ferritin level of below 2500 ng/mL.

12 Pulau Pinang

12.1 Introduction

Penang has a population of 1,774,000 as of 2019 (Department of Statistics Malaysia). Hospital Pulau Pinang (HPP) and Hospital Seberang Jaya (HSJ) are the only 2 hospitals providing care for all thalassaemia patients in the state.

12.2 Patient Demographics

Data analysed were taken from patients who are either alive, lost to follow-up or cured by transplant, and excludes the deceased patients. There are 499 living thalassaemia patients in Pulau Pinang. Five patients were cured by transplant and 32 patients were lost to follow-up. One patient with HbE/β-thalassaemia succumbed to renal failure in 2019.

Table 12.1: Distribution of Patients in Pulau Pinang by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Pulau Pinang	264	52.91
Hospital Seberang Jaya	235	47.09
Total	499	100.00

Table 12.2: Distribution of Patients in Pulau Pinang by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	462
Cured by Stem Cell Therapy	5
Total	467
Lost to Follow-up	32
Total	499
Deaths in 2019	1
Cumulative Reported Deaths	15

Table 12.3: Cumulative Causes of Death in Pulau Pinang Since 2007

Causes of Death	Number of Patients	Percentage (%)
Infections	7	46.67
Cardiac Causes	2	13.33
Tumour	1	6.67
Motor Vehicle Accident	1	6.67
Renal Failure	2	13.33
Central Nervous System Event	2	13.33
Total	15	100.00

12.2.1 Age Group

The patient in Pulau Pinang is 3 months old (diagnosed with β-thalassaemia major) and the eldest is 86 years old (HbH disease). A total of 217 patients (43.49%) are below 20 years old. There are 21 new thalassaemia births between 2014-2019, although with reducing trend.

Figure 12.1: Distribution of Patients in Pulau Pinang by Age Group

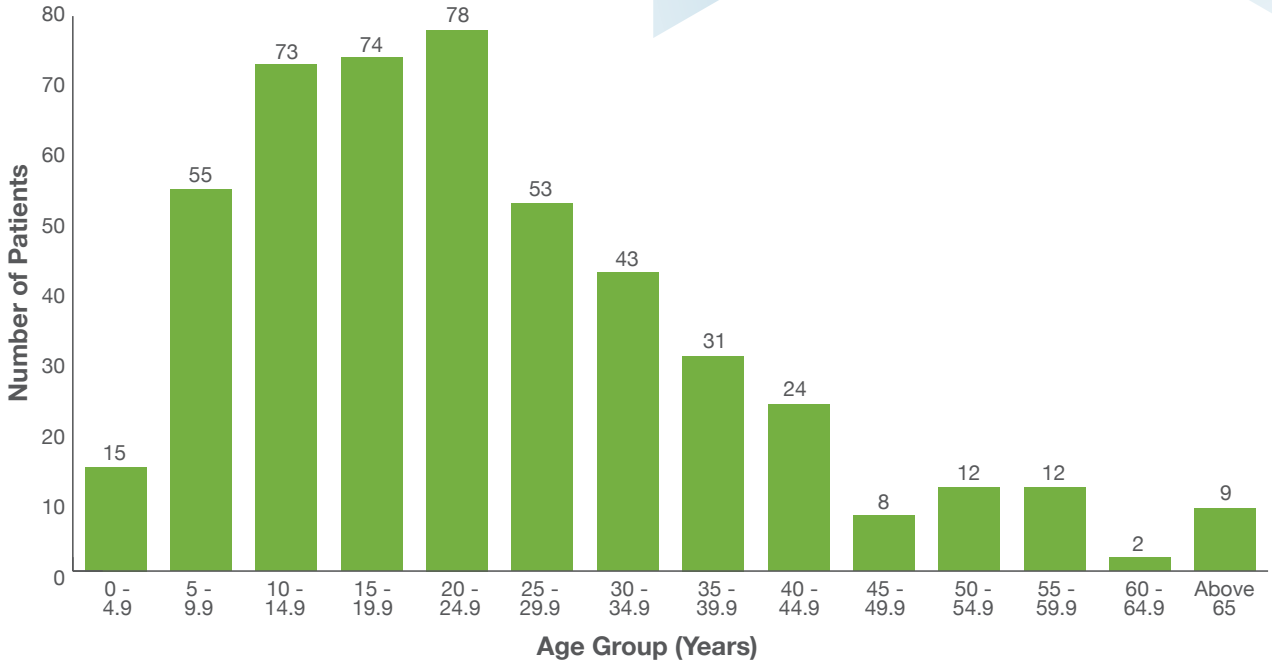


Table 12.4: Distribution of Patients in Pulau Pinang According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	143	β-Thalassaemia Major	30	20.98
		β-Thalassaemia Intermedia	11	7.69
		HbE/β-Thalassaemia	61	42.66
		HbH Disease	34	23.78
		Others	7	4.90
15 - 29.9	205	β-Thalassaemia Major	57	27.80
		β-Thalassaemia Intermedia	10	4.88
		HbE/β-Thalassaemia	85	41.46
		HbH Disease	46	22.44
		Others	7	3.41
30 - 44.9	98	β-Thalassaemia Major	18	18.37
		β-Thalassaemia Intermedia	11	11.22
		HbE/β-Thalassaemia	41	41.84
		HbH Disease	21	21.43
		Others	7	7.14
45 - 59.9	42	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	4	9.52
		HbE/β-Thalassaemia	12	28.57
		HbH Disease	25	59.52
		Others	1	2.38

60 and above	11	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	3	27.27
		HbH Disease	8	72.73
		Other	0	0.00
Total			499	

12.2.2 Gender

There are 222 male patients (44.49%) and 277 female patients (55.51%) in Pulau Pinang.

Table 12.5: Distribution of Patients in Pulau Pinang According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Pulau Pinang	115	23.05	149	29.86
Hospital Seberang Jaya	107	21.44	128	25.65
Total	222	44.49	277	55.51

12.2.3 Ethnic Group

Malay patients form the largest group of thalassaemia patients in Hospital Pulau Pinang and Hospital Seberang Jaya with 392 patients (78.56%), followed by the Chinese (89 patients, 17.84%), Indian (4 patients, 0.80%), and Kadazan-Dusun (3 patients, 0.60%). The remaining 11 patients (2.20%) are of Iban, Foreigner or Thai descent.

Figure 12.2: Distribution of Patients in Pulau Pinang by Ethnic Group

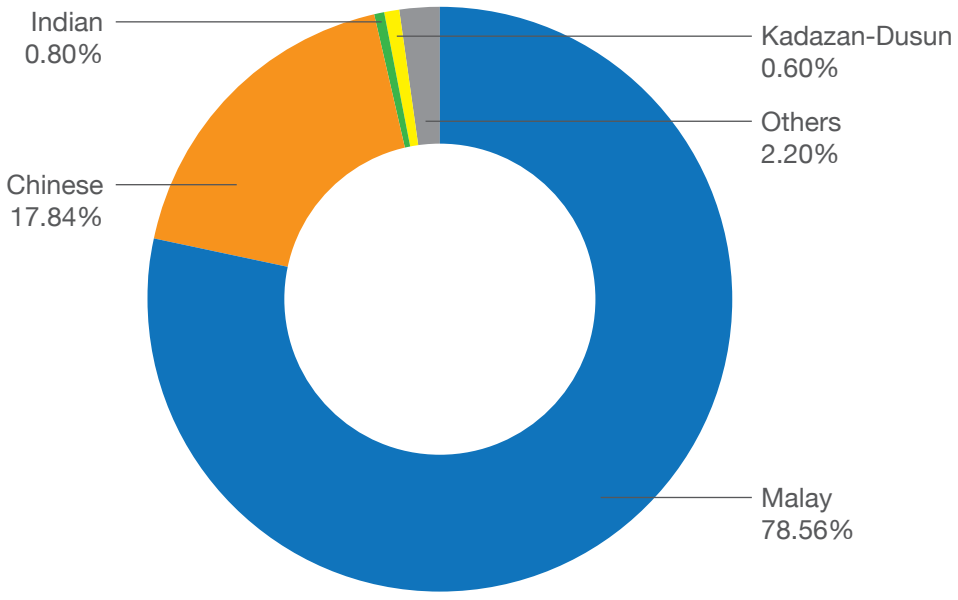


Table 12.6: Distribution of Patients in Pulau Pinang According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Others		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Pulau Pinang	180	36.07	73	14.63	1	0.20	3	0.60	6	1.20
Hospital Seberang Jaya	212	42.48	16	3.21	3	0.60	0	0.00	5	1.00
Total	392	78.56	89	17.84	4	0.80	3	0.60	11	2.20

12.3 Diagnosis

The most common diagnosis among patients in Pulau Pinang is HbE/β-thalassaemia with 201 patients (40.28%), followed by HbH disease (133 patients, 26.65%), β-thalassaemia major (105 patients, 21.04%), and β-thalassaemia intermedia (38 patients, 7.62%). The remaining 22 patients (4.41%) have other diagnoses, as shown in Figure 12.3.

Figure 12.3: Distribution of Patients in Pulau Pinang by Diagnosis

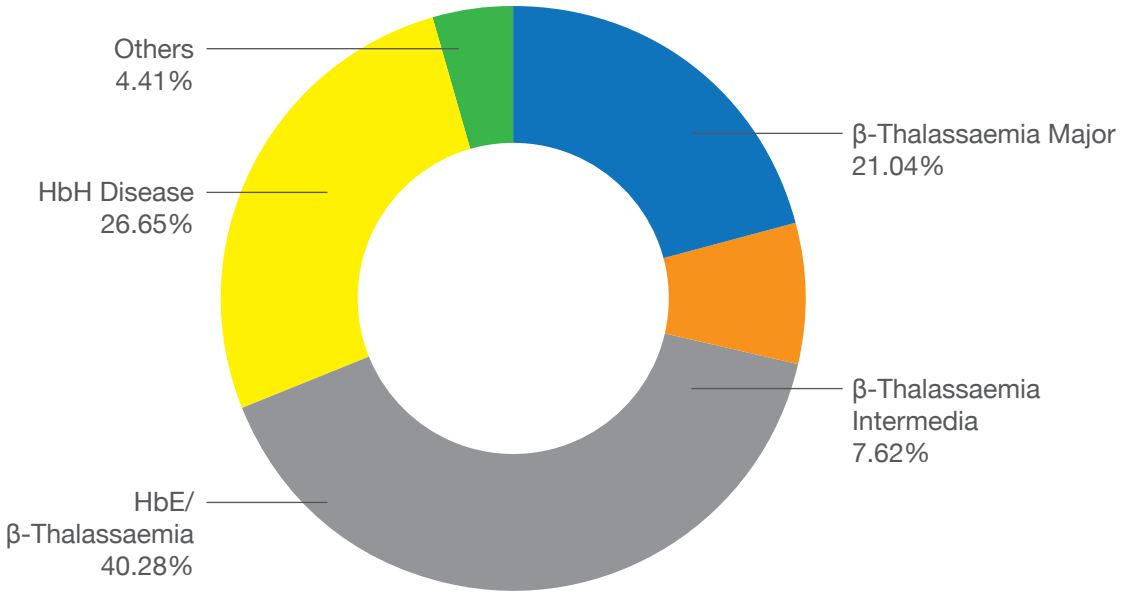


Table 12.7: Distribution of Patients in Pulau Pinang According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	105	Malay	73	14.63
		Chinese	28	5.61
		Indian	1	0.20
		Kadazan-Dusun	2	0.40
		Others	1	0.20
β-Thalassaemia Intermedia	38	Malay	28	5.61
		Chinese	6	1.20
		Indian	0	0.00
		Kadazan-Dusun	1	0.20
		Others	3	0.60
HbE/ β-Thalassaemia	201	Malay	183	36.67
		Chinese	13	2.61
		Indian	2	0.40
		Kadazan-Dusun	0	0.00
		Others	3	0.60

HbH Disease	133	Malay	95	19.04
		Chinese	35	7.01
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	3	0.60
Others*	22	Malay	14	2.81
		Chinese	7	1.40
		Indian	1	0.20
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Total			499	100.00

12.4 Treatment

12.4.1 Iron Chelation Therapy

A total of 317 patients (63.53%) in Pulau Pinang receive iron chelation therapy. Ninety-eight patients (30.91%) are on DFP, 77 patients (24.29%) are on DFX, 58 patients (18.30%) are on DFO, 66 patients (20.82%) are on a combination of DFO + DFP, 12 patients (3.79%) are on DFO + DFX and 6 patients (1.89%) are on DFP + DFX.

Table 12.8: Distribution of Patients in Pulau Pinang by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	58	18.30
DFP only	98	30.91
DFX only	77	24.29
DFO + DFP	66	20.82
DFP + DFX	6	1.89
DFO + DFX	12	3.79
DFO + DFP + DFX	0	0.00
Total	317	100.00

Table 12.9: Distribution of Patients in Pulau Pinang According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Pulau Pinang	144	DFO only	20	6.31
		DFP only	38	11.99
		DFX only	41	12.93
		DFO + DFP	38	11.99
		DFP + DFX	3	0.95
		DFO + DFX	4	1.26
		DFO + DFP + DFX	0	0.00
Hospital Seberang Jaya	173	DFO only	38	11.99
		DFP only	60	18.93
		DFX only	36	11.36
		DFO + DFP	28	8.83
		DFP + DFX	3	0.95
		DFO + DFX	8	2.52
		DFO + DFP + DFX	0	0.00
Total			317	100.00

Table 12.10 Distribution of Patients in Pulau Pinang According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	85	DFO only	15	17.65
		DFP only	14	16.47
		DFX only	41	48.24
		DFO + DFP	9	10.59
		DFP + DFX	2	2.35
		DFO + DFX	4	4.71
		DFO + DFP + DFX	0	0.00
15 - 29.9	138	DFO only	22	15.94
		DFP only	37	26.81
		DFX only	32	23.19
		DFO + DFP	39	28.26
		DFP + DFX	3	2.17
		DFO + DFX	5	3.62
		DFO + DFP + DFX	0	0.00
30 - 44.9	65	DFO only	11	16.92
		DFP only	36	55.38
		DFX only	2	3.08
		DFO + DFP	14	21.54
		DFP + DFX	1	1.54
		DFO + DFX	1	1.54
		DFO + DFP + DFX	0	0.00

45 - 59.9	27	DFO only	8	29.63
		DFP only	11	40.74
		DFX only	2	7.41
		DFO + DFP	4	14.81
		DFP + DFX	0	0.00
		DFO + DFX	2	7.41
		DFO + DFP + DFX	0	0.00
60 and above	2	DFO only	2	100.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			317	

12.4.2 Serum Ferritin Level

There are 224 regularly transfused patients in Pulau Pinang who had their serum ferritin levels measured in 2019. Of these, 42 patients (18.75%) have serum ferritin levels lower than 1000 ng/mL, 86 patients (38.39%) have serum ferritin levels between 1000 and 2500 ng/mL, and 96 patients (42.86%) have serum ferritin levels above 2500 ng/mL.

Table 12.11: Distribution of Patients in Pulau Pinang According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No	%	No	%	No	%	No	%	No	%
Hospital Pulau Pinang	115	18	8.04	51	22.77	24	10.71	18	8.04	4	1.79
Hospital Seberang Jaya	109	24	10.71	35	15.63	34	15.18	15	6.70	1	0.45
Total	224	42	18.75	86	38.39	58	25.89	33	14.73	5	2.23

12.5 Conclusion

HbE/β-thalassaemia is the most common type of thalassaemia in Pulau Pinang. The median age group of the patients is between 21-25 years old. The ethnic distribution of thalassaemia patients in Pulau Pinang is mainly Malay (78.56%), followed by Chinese (17.84%) and Indian (0.80%). The most common cause of death among thalassaemia patients in Pulau Pinang is infection, afflicting 7 out of 15 deceased patients. There are 5 patients with hepatitis C in Pulau Pinang, and 1 of them developed hepatocellular carcinoma.

DFP is the most common iron chelator used by patients in Pulau Pinang (98 patients, 30.91%), followed by DFX (77 patients, 24.29%), and DFO (58 patients, 18.30%). Among the TDT patients, 18.75% achieve serum ferritin levels below 1000 ng/mL, and 57.14% have serum ferritin levels below 2500 ng/mL.

13 Sabah

13.1 Introduction

Sabah is the second largest state in Malaysia after Sarawak, with a land area of 73.619 km^2. The population of Sabah in 2010 was reported at 3,206,742 (Department of Statistics Malaysia).

The population of Sabah consists of 32 officially recognised ethnic groups; 28 of these ethnic groups are considered as natives by the Federal Government of Malaysia. These natives are referred to as 'Bumiputera' or 'Pribumi'. The largest 'Bumiputera' ethnic group is the Kadazan-Dusun, followed by Bajau, Brunei and Murut. The non-'Bumiputera" ethnicities in Sabah are the Han Chinese, Indian and other races.

Table 13.1: Ethnic Distribution of Sabah Population

Ethnicity	Percentage (%)
Kadazan-Dusun	17.8
Malay*	5.71
Bajau	14
Murut	3.22
Other Bumiputera**	20.56
Chinese	9.11
Indian & Other Non-Bumiputera	1.5
Non-Malaysians	27.81

**Malay consists of Bruneian Malay, Kedayan, Banjar, Cocos and Peninsular Malay .*
***Consists of Rungus, Iranun, Bisaya, Suluk, Tagal, Sungai, Tidong, Bugis, Tatana, Lun Bawang/Lun Dayeh, Tindal, Tobilung, Kimaragang, Timogun, Nabay, Kagayan, Tombonuo, Sino, Jawa, Idahan, Makiang, Minokok, Lobu, Bonggi, Begahak, Talantang, Tinagas, Gana, Kuijau and others.*

The state of Sabah is divided into 5 administrative divisions, which in turn are sub-divided into 25 districts, as shown in Table 13.2.

Table 13.2: The Five Administrative Divisions of Sabah and Prevalence of Thalassaemia Patients in These Divisions

Division Name		Districts	Area (km^2)	Population in 2010	Number of Patients	Prevalence (per 100,000 Population)
1	Pantai Barat (Kota Kinabalu)	Kota Belud, Kota Kinabalu, Papar, Penampang, Putatan, Ranau, Tuaran	7,588	1,067,589	874	81.87
2	Pedalaman (Keningau)	Beaufort, Keningau, Nabawan, Kuala Penyu, Sipitang, Tambunan, Tenom	18,298	424,534	312	73.49
3	Kudat	Kota Marudu, Kudat, Pitas	4,623	192,457	286	148.60
4	Sandakan	Beluran, Kinabatangan, Sandakan, Tongod	28,205	702,207	239	34.02
5	Tawau	Kunak, Lahad Datu, Semporna, Tawau	14,905	819,955	103	12.56
Total			-	3,206,742	1814	-

13.2 Patient Demographics

There are 1829 thalassaemia patients in Sabah registered in the MTR until November 2019.

Table 13.3: Distribution of Thalassaemia Patients and Estimated Prevalence Rate in Each Treatment Centre in Sabah

Centre	Number of Patients	Percentage (%)	Population in 2010#	Prevalence Rate (per 100,000 population)
Hospital Wanita & Kanak-Kanak Sabah, Likas*	307	16.79	750,175	40.92
Hospital Queen Elizabeth, Kota Kinabalu*	369	20.17		
Hospital Tuaran	18	0.98		
Hospital Duchess of Kent, Sandakan	197	10.77	409,056	48.15
Hospital Keningau^	179	9.79	210,044	85.22
Hospital Kota Marudu	135	7.38	68,289	197.68
Hospital Ranau	98	5.36	95,800	102.29
Hospital Kudat	71	3.88	85,404	83.13
Hospital Pitas	85	4.65	38,764	219.27
Hospital Kota Belud	43	2.35	93,180	46.14
Hospital Papar	46	2.52	128,434	35.81
Hospital Tawau	53	2.90	412,375	12.85
Hospital Tenom	25	1.37	56,597	44.17
Hospital Tambunan	36	1.97	36,297	99.18
Hospital Lahad Datu	41	2.24	206,861	19.18
Hospital Kinabatangan**	29	1.59	186,519	15.55
Hospital Beaufort	30	1.64	66,406	45.18
Hospital Beluran	18	0.98	106,632	16.88
Hospital Kuala Penyu	18	0.98	19,426	92.66
Hospital Sipitang	22	1.20	35,764	61.90
Hospital Kunak	7	0.38	62,851	11.14
Hospital Semporna	2	0.11	137,868	1.45
Total	1829	100.00	3,206,742	

 # Department of Statistics Malaysia, census July 2010.
**Total population of Kota Kinabalu, Penampang, Putatan and Tuaran.*
^Total population of Keningau and Nabawan.
***Total population of Kinabatangan and Tongod.*

The 3 hospitals with the highest number of patients in Sabah are Hospital Queen Elizabeth (HQE), Hospital Wanita dan Kanak-Kanak Sabah (HWKKS) and Hospital Duchess of Kent (HDOK).

The pattern of distribution of thalassaemia patients in Sabah follows closely the distribution of its ethnic groups, which have a high carrier rate of β-thalassaemia among the Kadazan-Dusun, Rungus, Bajau and Murut. This explains why small districts such as Keningau, Kota Marudu, Pitas and Ranau have a large number of thalassaemia patients. The bigger hospitals in the southeast of Sabah, e.g. Hospital Tawau and Hospital Lahad Datu, have relatively fewer patients, as there is relatively low density of Kadazan-Dusun, Rungus, Bajau and Murut populations in these towns.

Figure 13.1: Distribution of Patients in Sabah by Centre

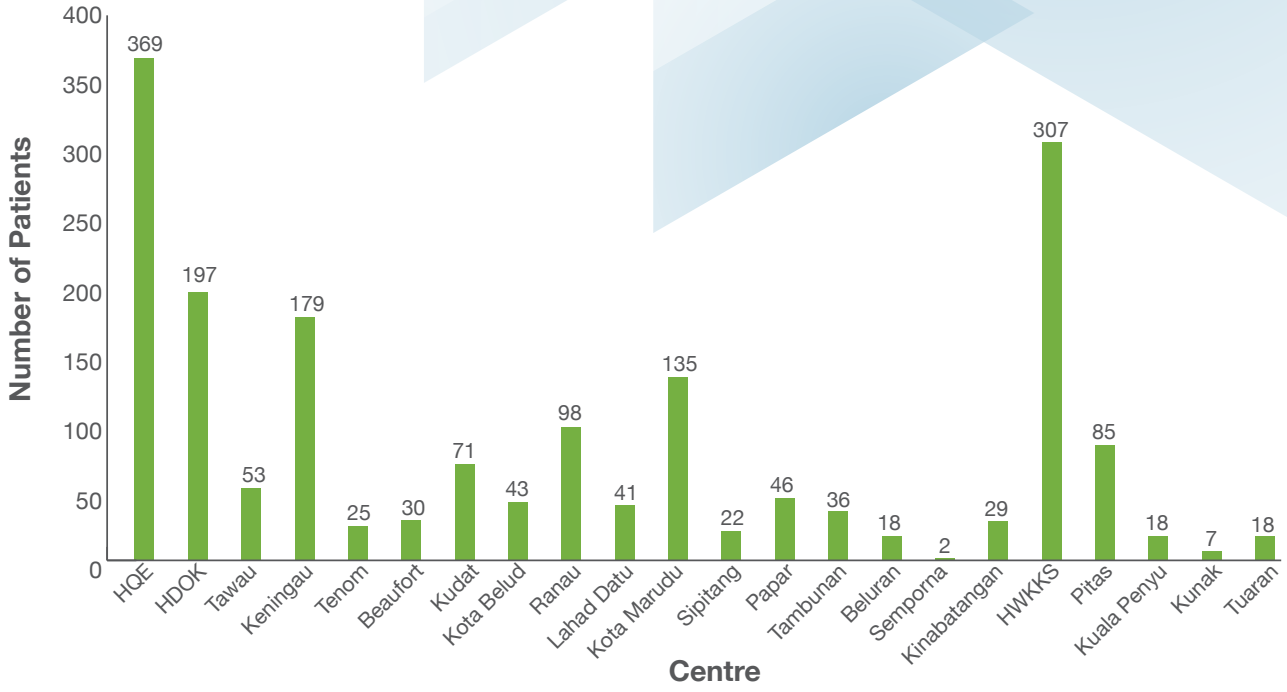


Table 13.4: Distribution of Patients in Sabah by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	1690
Cured by Stem Cell Therapy (2008-2019)	37
Total	1727
Lost to Follow-up	102
Total	1829
Deaths in 2019	14
Cumulative Reported Deaths	319

Table 13.5: Cumulative Causes of Death in Sabah Since 2007

Causes of Death	Number of Patients	Percentage
Cardiac Causes	117	47.37
Endocrine Complications	5	2.02
Infections	88	35.63
Central Nervous System Event	5	2.02
Arrhythmias	2	0.81
Thrombosis	2	0.81
Liver Disease	5	2.02
Motor Vehicle Accidents	4	1.62
Renal Complications	2	0.81
Surgical Complications	3	1.21
Thalassaemia	3	1.21
Tumours	4	1.62
Others	7	2.83
Total	247	100.0

There were 14 deaths reported in 2019, showing reducing trend compared to 18 deaths reported in 2018. Seventy-two of the deaths in Sabah had an unknown cause of death due to unavailable data.

13.2.1 Age Group

In the current 2019 statistics, the age group with the highest number of thalassaemia patients in Sabah is 10-14.9 years. This is in contrast with the data in 2017, in which the age group with highest number of thalassaemia patients was 6-10 years. This reflects an improvement in patients' survival rates. The oldest surviving thalassaemia major patient in Sabah is currently 50 years old.

In 2019, the number of thalassaemia patients younger than 15 years old continue to increase in number, compared to in 2018. However, the overall percentage of patients younger than 15 years old in 2019 had reduced in view of the increasing survival rates of whole cohorts of thalassaemia patients.

Figure 13.2: Distribution of Patients in Sabah by Age Group

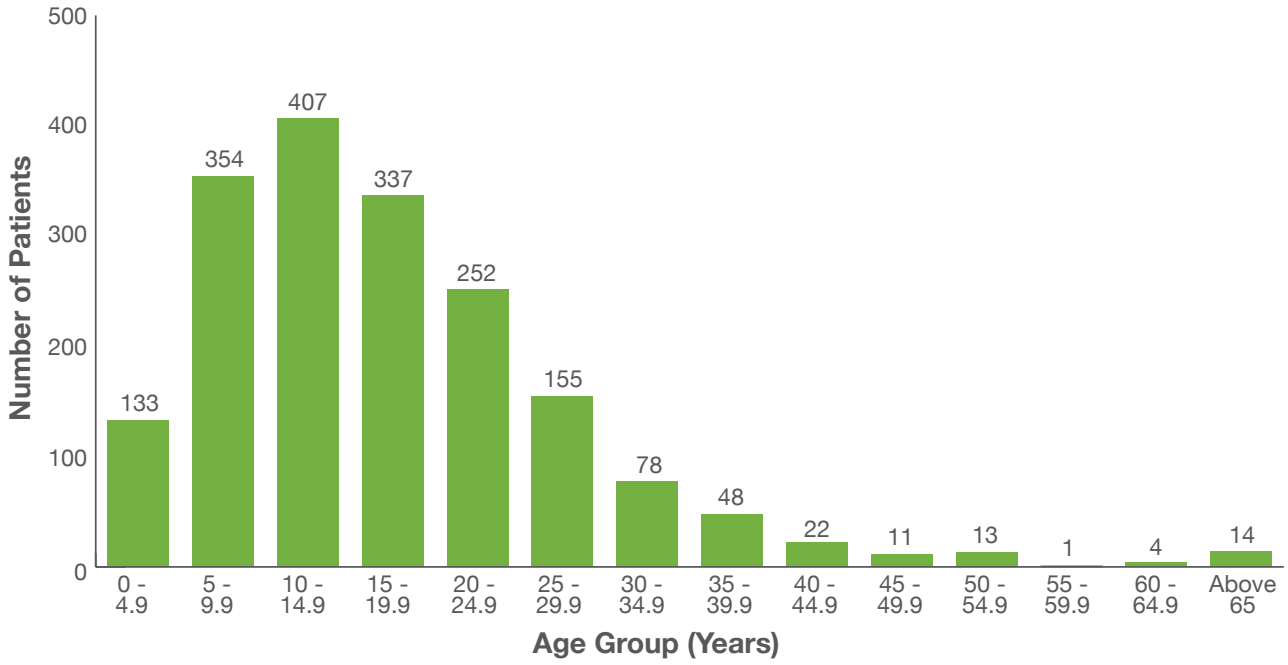


Table 13.6: Distribution of Patients in Sabah According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	894	β-Thalassaemia Major	684	76.51
		β-Thalassaemia Intermedia	106	11.86
		HbE/β-Thalassaemia	65	7.27
		HbH Disease	35	3.91
		Others	4	0.45
15 - 29.9	744	β-Thalassaemia Major	575	77.28
		β-Thalassaemia Intermedia	116	15.59
		HbE/β-Thalassaemia	32	4.30
		HbH Disease	21	2.82
		Others	0	0.00
30 - 44.9	148	β-Thalassaemia Major	60	40.54
		β-Thalassaemia Intermedia	45	30.41
		HbE/β-Thalassaemia	19	12.84
		HbH Disease	23	15.54
		Others	1	0.68
45 - 59.9	25	β-Thalassaemia Major	2	8.00
		β-Thalassaemia Intermedia	5	20.00
		HbE/β-Thalassaemia	11	44.00
		HbH Disease	7	28.00
		Others	0	0.00
60 and above	18	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	11.11
		HbE/β-Thalassaemia	3	16.67
		HbH Disease	13	72.22
		Others	0	0.00
Total			1829	

β-thalassaemia major constitutes the majority diagnosis in Sabah. Up to early November 2019, 72.22% of the total number of patients were diagnosed with β-thalassaemia major, and 15% were diagnosed with β-thalassaemia intermedia. HbE/β-thalassaemia constitutes much fewer number of cases in Sabah compared to West Malaysia. This observation is consistent with the discovery of the severe 45-kb β-globin gene deletion (Filipino deletion) occurring in homozygous form in more than 90% of Sabahan Kadazan-Dusun with β-thalassaemia syndrome.

In comparison with MTR data reported in year 2018, there was an increase of 15 thalassaemia patients in year 2019, including thalassaemia major patients. This increased number of patients impose increased workload and healthcare burden to the treatment centres in Sabah.

13.2.2 Gender

The gender distribution of thalassaemia patients in Sabah is nearly equal, i.e. 949 male patients (51.89%) and 880 female patients (48.11%), as is consistent with thalassaemia syndrome as an autosomal recessive disease.

Table 13.7: Distribution of Patients in Sabah According to Gender by Centre

Centre	Total Number of Patients	Male		Female	
		Number	%	Number	%
Hospital Queen Elizabeth, Kota Kinabalu	369	167	9.13	202	11.04
Hospital Duchess of Kent, Sandakan	197	100	5.47	97	5.30
Hospital Tawau	53	24	1.31	29	1.59
Hospital Keningau	179	91	4.98	88	4.81
Hospital Tenom	25	11	0.60	14	0.77
Hospital Beaufort	30	18	0.98	12	0.66
Hospital Kudat	71	39	2.13	32	1.75
Hospital Kota Belud	43	26	1.42	17	0.93
Hospital Ranau	98	59	3.23	39	2.13
Hospital Lahad Datu	41	15	0.82	26	1.42
Hospital Kota Marudu	135	69	3.77	66	3.61
Hospital Sipitang	22	14	0.77	8	0.44
Hospital Papar	46	33	1.80	13	0.71
Hospital Tambunan	36	22	1.20	14	0.77
Hospital Beluran	18	12	0.66	6	0.33
Hospital Semporna	2	0	0.00	2	0.11
Hospital Kinabatangan	29	15	0.82	14	0.77
Hospital Hospital Wanita & Kanak-Kanak Sabah, Likas	307	158	8.64	149	8.15
Hospital Pitas	85	44	2.41	41	2.24
Hospital Kuala Penyu	18	13	0.71	5	0.27
Hospital Kunak	7	5	0.27	2	0.11
Hospital Tuaran	18	14	0.77	4	0.22
Total	1829	949	51.89	880	48.11

13.2.3 Ethnic Group

The Kadazan-Dusun form the largest group of patients in Sabah with 862 patients (47.13%), followed by Pribumi Sabah with 211 patients (11.54%).

Figure 13.3: Distribution of Patients in Sabah by Ethnic Group

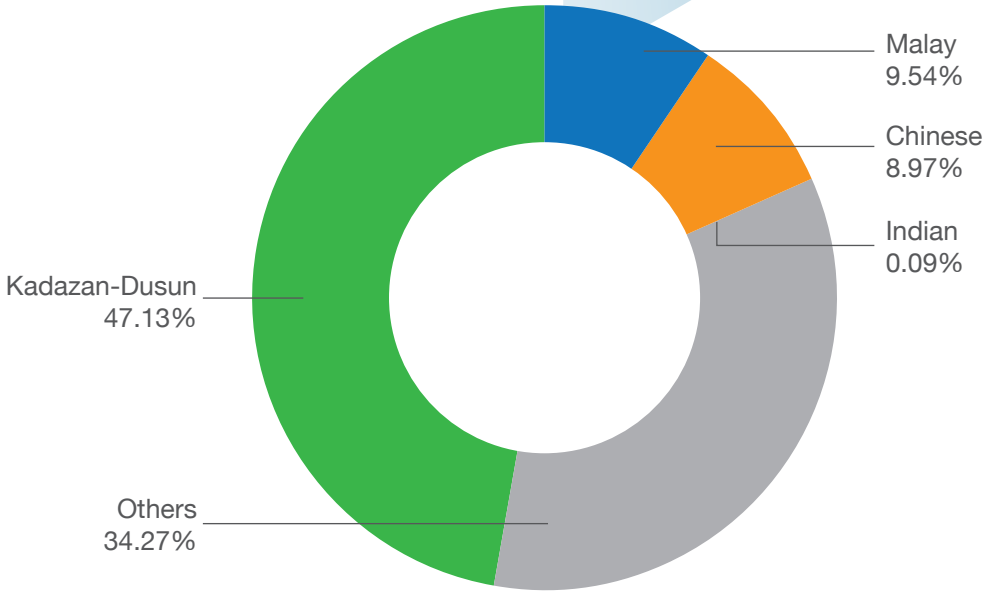


Figure 13.4: Distribution of Patients in Sabah by Ethnic Sub-Group Other Than Malay, Chinese, Indian and Kadazan-Dusun

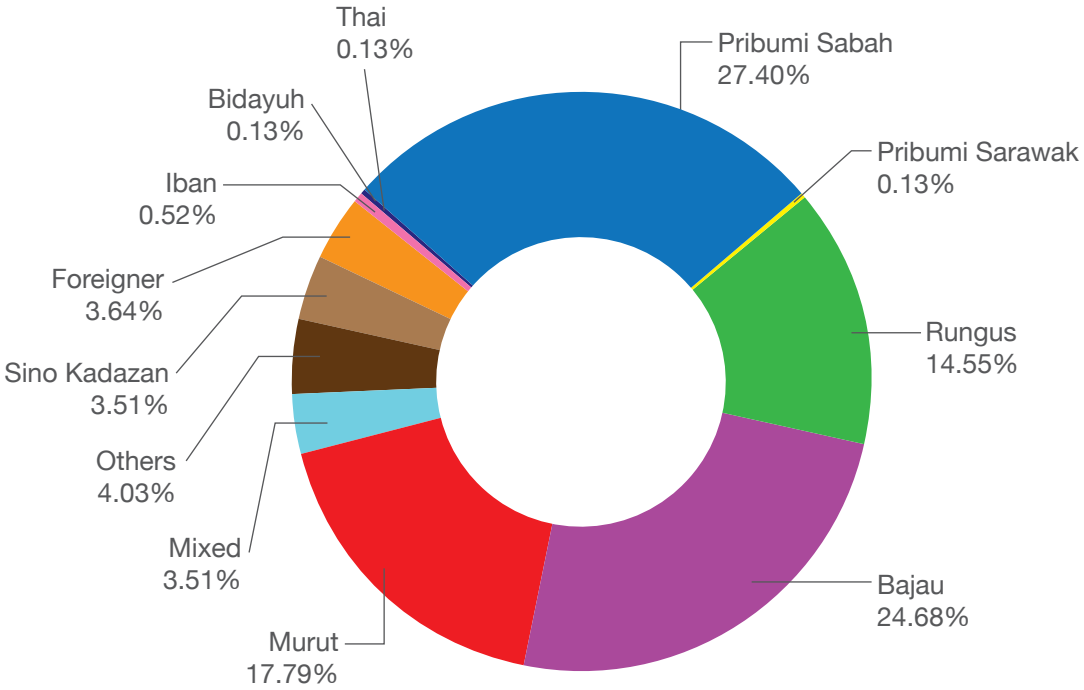


Table 13.8: Distribution of Patients in Sabah According to Ethnic Group by Centre

Ethnic Group	Kadazan-Dusun		Rungus		Malay		Bajau		Murut		Chinese		Other	
Centre	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
HQE	165	9.02	30	1.64	26	1.42	56	3.06	9	0.49	47	2.57	10	0.55
HDOK	54	2.95	0	0.00	17	0.93	26	1.42	0	0.00	7	0.38	11	0.60
Tawau	8	0.44	0	0.00	6	0.33	13	0.71	0	0.00	7	0.38	1	0.05
Keningau	88	4.81	0	0.00	1	0.05	1	0.05	80	4.37	5	0.27	0	0.00
Tenom	2	0.11	0	0.00	2	0.11	1	0.05	17	0.93	2	0.11	0	0.00
Beaufort	6	0.33	0	0.00	4	0.22	2	0.11	2	0.11	0	0.00	0	0.00
Kudat	8	0.44	42	2.30	0	0.00	7	0.38	0	0.00	4	0.22	4	0.22
Kota Belud	35	1.91	0	0.00	1	0.05	4	0.22	1	0.05	0	0.00	0	0.00
Ranau	98	5.36	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Lahad Datu	5	0.27	0	0.00	4	0.22	15	0.82	0	0.00	1	0.05	1	0.05
Kota Marudu	100	5.47	11	0.60	1	0.05	5	0.27	0	0.00	1	0.05	2	0.11
Sipitang	0	0.00	0	0.00	3	0.16	1	0.05	14	0.77	0	0.00	0	0.00
Papar	22	1.20	3	0.16	7	0.38	5	0.27	3	0.16	0	0.00	0	0.00
Tambunan	32	1.75	0	0.00	0	0.00	0	0.00	2	0.11	0	0.00	0	0.00
Beluran	14	0.77	0	0.00	0	0.00	1	0.05	0	0.00	0	0.00	0	0.00
Semporna	0	0.00	0	0.00	0	0.00	2	0.11	0	0.00	0	0.00	0	0.00
Kinabatangan	1	0.05	0	0.00	3	0.16	5	0.27	0	0.00	0	0.00	1	0.05
HWKKS	158	8.64	11	0.60	23	1.26	35	1.91	9	0.49	19	1.04	0	0.00
Pitas	39	2.13	15	0.82	2	0.11	2	0.11	0	0.00	1	0.05	1	0.05
Kuala Penyu	11	0.60	0	0.00	1	0.05	3	0.16	0	0.00	0	0.00	0	0.00
Kunak	0	0.00	0	0.00	0	0.00	5	0.27	0	0.00	0	0.00	0	0.00
Tuaran	16	0.87	0	0.00	0	0.00	1	0.05	0	0.00	1	0.05	0	0.00
Total	862	47.13	112	6.12	101	5.52	190	10.39	137	7.49	95	5.19	31	1.69

A significant percentage of thalassaemia patients in Sabah are of Kadazan-Dusun descent (47.13% of total number of patients). This is disproportionately high compared to the percentage of Kadazan-Dusun in the population of Sabah. The prevalence of thalassaemia cases are also disproportionately high among the Bajau (10.39%) and Murut (7.49%) ethnic groups.

The predominance of thalassaemia syndrome among the Kadazan-Dusun and in the west coast of Sabah has implications on the allocation of resources towards effective thalassaemia control.

13.3 Diagnosis

β-thalassaemia major constitutes the majority diagnosis in Sabah. Up to early November 2019, 72.23% of the thalassaemia patients in Sabah were diagnosed with β-thalassaemia major, and 14.98% were diagnosed with β-thalassaemia intermedia. HbE/β-thalassaemia constitutes much fewer number of cases in Sabah as compared to West Malaysia. This observation is consistent with the discovery of a severe mutation, i.e. a 45-kb β-globin gene deletion occurring in homozygous form in more than 90% of Sabahan Kadazan-Dusun with β-thalassaemia syndrome.

Figure 13.5: Distribution of Patients in Sabah by Diagnosis

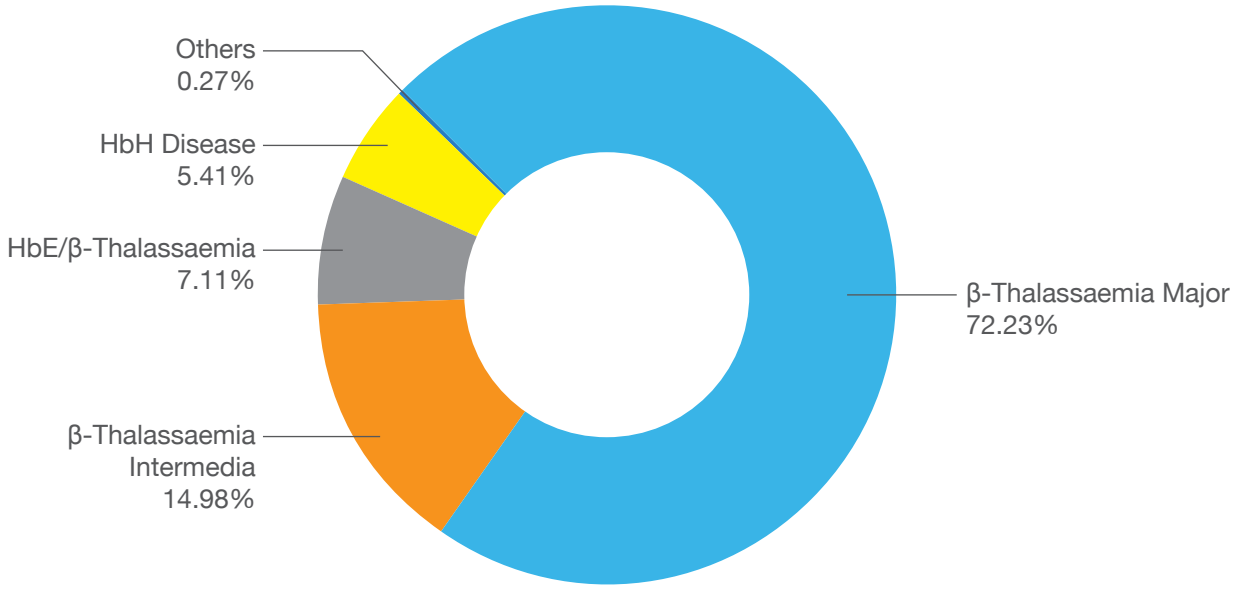


Table 13.9: Distribution of Patients in Sabah According to Diagnosis by Centre

Centre	Diagnosis									
	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/ β-Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Queen Elizabeth, Kota Kinabalu	220	12.03	60	3.28	39	2.13	49	2.68	1	0.05
Hospital Duchess of Kent, Sandakan	113	6.18	47	2.57	29	1.59	8	0.44	0	0.00
Hospital Tawau	26	1.42	14	0.77	6	0.33	7	0.38	0	0.00
Hospital Keningau	164	8.97	11	0.60	2	0.11	1	0.05	1	0.05
Hospital Tenom	22	1.20	1	0.05	1	0.05	1	0.05	0	0.00
Hospital Beaufort	23	1.26	7	0.38	0	0.00	0	0.00	0	0.00
Hospital Kudat	53	2.90	5	0.27	5	0.27	8	0.44	0	0.00
Hospital Kota Belud	34	1.86	4	0.22	5	0.27	0	0.00	0	0.00
Hospital Ranau	88	4.81	7	0.38	2	0.11	0	0.00	1	0.05
Hospital Lahad Datu	23	1.26	9	0.49	5	0.27	4	0.22	0	0.00
Hospital Kota Marudu	112	6.12	16	0.87	5	0.27	2	0.11	0	0.00
Hospital Sipitang	18	0.98	3	0.16	0	0.00	1	0.05	0	0.00
Hospital Papar	38	2.08	6	0.33	2	0.11	0	0.00	0	0.00
Hospital Tambunan	34	1.86	2	0.11	0	0.00	0	0.00	0	0.00
Hospital Beluran	18	0.98	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Semporna	2	0.11	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kinabatangan	24	1.31	4	0.22	1	0.05	0	0.00	0	0.00
Hospital Wanita & Kanak-Kanak Sabah, Likas	206	11.26	61	3.34	22	1.20	17	0.93	1	0.05
Hospital Pitas	73	3.99	5	0.27	6	0.33	1	0.05	0	0.00
Hospital Kuala Penyu	11	0.60	6	0.33	0	0.00	0	0.00	1	0.05
Hospital Kunak	2	0.11	5	0.27	0	0.00	0	0.00	0	0.00
Hospital Tuaran	17	0.93	1	0.05	0	0.00	0	0.00	0	0.00
Total	**1321**	**72.23**	**274**	**14.98**	**130**	**7.11**	**99**	**5.41**	**5**	**0.27**

Table 13.10: Distribution of Patients in Sabah According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	1321	Malay	54	2.95
		Chinese	45	2.46
		Indian	1	0.05
		Kadazan-Dusun	742	40.57
		Others:		
		Rungus	97	5.3
		Bajau	77	4.21
		Murut	121	6.62
		Mixed	14	0.77
		Others	170	9.29
β-Thalassaemia Intermedia	274	Malay	22	1.2
		Chinese	8	0.44
		Indian	0	0
		Kadazan-Dusun	85	4.65
		Others:		
		Rungus	12	0.66
		Bajau	48	2.62
		Murut	13	0.71
		Mixed	9	0.49
		Others	77	4.21
HbE/ β-Thalassaemia	130	Malay	17	0.93
		Chinese	6	0.33
		Indian	0	0
		Kadazan-Dusun	23	1.26
		Others:		
		Rungus	1	0.05
		Bajau	42	2.3
		Murut	0	0
		Mixed	4	0.22
		Others	37	2.02
HbH Disease	99	Malay	7	0.38
		Chinese	36	1.97
		Indian	0	0
		Kadazan-Dusun	10	0.55
		Others		
		Rungus	2	0.11
		Bajau	21	1.15
		Murut	3	0.16
		Mixed	0	0
		Others	20	1.09

Others	5	Malay	1	0.05
		Chinese	0	0
		Indian	0	0
		Kadazan-Dusun	2	0.11
		Others:		
		Rungus	0	0
		Bajau	2	0.11
		Murut	0	0
		Mixed	0	0
		Others	0	0
Total			1829	100.00

13.4 Treatment

13.4.1 Iron Chelation Therapy

The distribution patients in Sabah according to type of iron chelator received are shown in Table 13.11.

Table 13.11: Distribution of Patients in Sabah by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	327	26.63
DFP only	155	12.62
DFX only	337	27.44
DFO + DFP	270	21.99
DFP + DFX	48	3.91
DFO + DFX	60	4.89
DFO + DFP + DFX	31	2.52
Total	1228	100.00

DFX is the most common type of iron chelator prescribed (27.44%), followed by DFO (26.63%) and a combination of DFO and DFP (21.99%). DFX is mainly prescribed to paediatric patients. Many older thalassaemia patients in Sabah started chelation therapy later in life, i.e. when the government began to provide free chelation treatment in 2006. Many of these patients already had severe iron overload; hence these patients are treated with a combination of DFO and DFP. A total of 601 patients in Sabah were registered as 'Not on Iron Chelation Therapy'. However, the figure is likely to be inaccurate, caused by incomplete data entry or technical errors and requires further verification.

Table 13.12: Distribution of Patients in Sabah According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Queen Elizabeth, Kota Kinabalu	241	DFO only	112	9.12
		DFP only	26	2.12
		DFX only	2	0.16
		DFO + DFP	78	6.35
		DFP + DFX	1	0.08
		DFO + DFX	5	0.41
		DFO + DFP + DFX	17	1.38

Hospital Duchess of Kent, Sandakan	149	DFO only	54	4.40
		DFP only	20	1.63
		DFX only	38	3.09
		DFO + DFP	25	2.04
		DFP + DFX	4	0.33
		DFO + DFX	7	0.57
		DFO + DFP + DFX	1	0.08
Hospital Tawau	38	DFO only	2	0.16
		DFP only	4	0.33
		DFX only	13	1.06
		DFO + DFP	9	0.73
		DFP + DFX	4	0.33
		DFO + DFX	5	0.41
		DFO + DFP + DFX	1	0.08
Hospital Keningau	149	DFO only	13	1.06
		DFP only	29	2.36
		DFX only	30	2.44
		DFO + DFP	47	3.83
		DFP + DFX	20	1.63
		DFO + DFX	7	0.57
		DFO + DFP + DFX	3	0.24
Hospital Tenom	20	DFO only	6	0.49
		DFP only	2	0.16
		DFX only	3	0.24
		DFO + DFP	7	0.57
		DFP + DFX	1	0.08
		DFO + DFX	1	0.08
		DFO + DFP + DFX	0	0.00
Hospital Beaufort	23	DFO only	7	0.57
		DFP only	3	0.24
		DFX only	10	0.81
		DFO + DFP	2	0.16
		DFP + DFX	0	0.00
		DFO + DFX	1	0.08
		DFO + DFP + DFX	0	0.00
Hospital Kudat	55	DFO only	9	0.73
		DFP only	2	0.16
		DFX only	21	1.71
		DFO + DFP	19	1.55
		DFP + DFX	0	0.00
		DFO + DFX	3	0.24
		DFO + DFP + DFX	1	0.08

Hospital Kota Belud	21	DFO only	9	0.73
		DFP only	1	0.08
		DFX only	10	0.81
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.08
		DFO + DFP + DFX	0	0.00
Hospital Ranau	59	DFO only	32	2.61
		DFP only	1	0.08
		DFX only	18	1.47
		DFO + DFP	6	0.49
		DFP + DFX	0	0.00
		DFO + DFX	1	0.08
		DFO + DFP + DFX	1	0.08
Hospital Lahad Datu	28	DFO only	7	0.57
		DFP only	3	0.24
		DFX only	10	0.81
		DFO + DFP	5	0.41
		DFP + DFX	1	0.08
		DFO + DFX	1	0.08
		DFO + DFP + DFX	1	0.08
Hospital Kota Marudu	99	DFO only	7	0.57
		DFP only	23	1.87
		DFX only	24	1.95
		DFO + DFP	40	3.26
		DFP + DFX	2	0.16
		DFO + DFX	3	0.24
		DFO + DFP + DFX	0	0.00
Hospital Sipitang	19	DFO only	2	0.16
		DFP only	2	0.16
		DFX only	9	0.73
		DFO + DFP	4	0.33
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	2	0.16
Hospital Papar	23	DFO only	4	0.33
		DFP only	6	0.49
		DFX only	11	0.90
		DFO + DFP	0	0.00
		DFP + DFX	2	0.16
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Tambunan	31	DFO only	5	0.41
		DFP only	2	0.16
		DFX only	11	0.90
		DFO + DFP	6	0.49
		DFP + DFX	3	0.24
		DFO + DFX	2	0.16
		DFO + DFP + DFX	2	0.16
Hospital Beluran	16	DFO only	3	0.24
		DFP only	3	0.24
		DFX only	5	0.41
		DFO + DFP	5	0.41
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Semporna	2	DFO only	0	0.00
		DFP only	1	0.08
		DFX only	1	0.08
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kinabatangan	25	DFO only	11	0.90
		DFP only	1	0.08
		DFX only	8	0.65
		DFO + DFP	2	0.16
		DFP + DFX	0	0.00
		DFO + DFX	2	0.16
		DFO + DFP + DFX	1	0.08
Hospital Wanita & Kanak-Kanak Sabah, Likas	164	DFO only	22	1.79
		DFP only	21	1.71
		DFX only	89	7.25
		DFO + DFP	8	0.65
		DFP + DFX	6	0.49
		DFO + DFX	17	1.38
		DFO + DFP + DFX	1	0.08
Hospital Pitas	36	DFO only	18	1.47
		DFP only	2	0.16
		DFX only	10	0.81
		DFO + DFP	3	0.24
		DFP + DFX	3	0.24
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Kuala Penyu	11	DFO only	2	0.16
		DFP only	1	0.08
		DFX only	2	0.16
		DFO + DFP	2	0.16
		DFP + DFX	0	0.00
		DFO + DFX	4	0.33
		DFO + DFP + DFX	0	0.00
Hospital Kunak	3	DFO only	0	0.00
		DFP only	2	0.16
		DFX only	0	0.00
		DFO + DFP	1	0.08
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tuaran	16	DFO only	2	0.16
		DFP only	0	0.00
		DFX only	12	0.98
		DFO + DFP	1	0.08
		DFP + DFX	1	0.08
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			1228	100.00

Table 13.13: Distribution of Patients in Sabah According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0–14.9	558	DFO only	64	11.47
		DFP only	62	11.11
		DFX only	310	55.56
		DFO + DFP	37	6.63
		DFP + DFX	38	6.81
		DFO + DFX	38	6.81
		DFO + DFP + DFX	9	1.61
15–29.9	582	DFO only	240	41.24
		DFP only	60	10.31
		DFX only	27	4.64
		DFO + DFP	208	35.74
		DFP + DFX	10	1.72
		DFO + DFX	20	3.44
		DFO + DFP + DFX	17	2.92
30–44.9	80	DFO only	21	26.25
		DFP only	27	33.75
		DFX only	0	0.00
		DFO + DFP	25	31.25
		DFP + DFX	0	0.00
		DFO + DFX	2	2.50
		DFO + DFP + DFX	5	6.25

45–59.9	8	DFO only	2	25.00
		DFP only	6	75.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			1228	

13.4.2 Serum Ferritin Level

From the limited data on serum ferritin level retrievable from the MTR up to November 2019, only 362 patients in Sabah had their serum ferritin level measured in 2019. A total of 82 patients (22.65%) have a serum ferritin level below 1000 ng/mL, 78 patients (21.55%) have a serum ferritin level between 1000-2499 ng/mL, and 87 patients (24.03%) have a serum ferritin level between 2500-4999 ng/mL. Eighty-six patients (23.76%) have a serum ferritin level between 5000-9999 ng/mL, and 29 patients (8.01%) have a serum ferritin level above 10000 ng/mL.

Table 13.14: Distribution of Patients in Sabah According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Beaufort	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Beluran	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
HDOK	120	21	5.80	30	8.29	33	9.12	34	9.39	2	0.55
HWKKS	17	5	1.38	5	1.38	2	0.55	3	0.83	2	0.55
Keningau	44	8	2.21	13	3.59	9	2.49	7	1.93	7	1.93
Kinabatangan	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Kota Belud	1	1	0.28	0	0.00	0	0.00	0	0.00	0	0.00
Kota Marudu	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Kuala Penyu	1	0	0.00	0	0.00	0	0.00	1	0.28	0	0.00
Kudat	14	4	1.10	3	0.83	5	1.38	2	0.55	0	0.00
Kunak	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Lahad Datu	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Papar	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Pitas	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
HQE	141	39	10.77	21	5.80	31	8.56	34	9.39	16	4.42
Ranau	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Semporna	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Sipitang	14	2	0.55	3	0.83	5	1.38	3	0.83	1	0.28
Tambunan	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00

Tawau	10	2	0.55	3	0.83	2	0.55	2	0.55	1	0.28
Tenom	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Tuaran	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	362	82	22.65	78	21.55	87	24.03	86	23.76	29	8.01

13.5 Conclusion

There are 16 new thalassaemia patients reported in Sabah in 2019. This is a reduction in the number of new cases in Sabah compared to 2018. Thalassaemia patients in Sabah are frequently treated at HQE (369 patients), followed by HWKKS (307 patients), HDOK (197 patients) and Hospital Keningau (179 patients). A majority of these patients are of Kadazan-Dusun ethnicity. β-thalassaemia major is the most common diagnosis in the state, leading to Sabah having the highest prevalence of the diagnosis in the country. The age group with the highest number of patients is 11-15 years old with 407 patients, followed 6-10 years old (354 patients) and 16-20 years old (337 patients). These results show that management of thalassaemia in paediatric patients are improving and sustained.

14 Sarawak

14.1 Introduction

The population of Sarawak in 2019 is 2,868,700 people (Department of Statistics Malaysia). Sarawak exhibits notable diversity in ethnicity, culture and language. There are 40 ethnic groups in Sarawak. The Iban form the largest group (29%), followed by Malay (23%), Chinese (22%), Bidayuh (8%), Melanau (5%) and other Bumiputera (7%). Non-citizens make up 6% of population.

Sarawak's main centre of treatment is Hospital Umum Sarawak. There are 5 other district hospitals in Sarawak, namely Hospital Miri, Hospital Sibu, Hospital Bintulu, Hospital Lawas and Hospital Limbang. All district hospitals have their own specialist-in-charge, except for Hospital Lawas.

Up to November 2019, there are 265 thalassaemia patients registered in MTR in Sarawak. Fourteen of these patients have been cured by stem cell transplant, and 21 patients have passed away since the registry was started in 1997. No thalassaemia deaths were recorded in 2019 (up to the end of October).

Table 14.1: Distribution of Thalassaemia Patients in Sarawak According to Thalassaemia Type by District

Administrative District	Number of β-Thalassaemia Patients	Number of α-Thalassaemia Patients	Total Population
Kuching Division	**90**	**44**	**802,900**
Kuching	87	44	702,700
Bau	1	0	61,500
Lundu	2	0	38,700
Samarahan Division	**1**	**9**	**184,800**
Samarahan	1	8	101,300
Simunjan	0	1	46,200
Asajaya	0	0	37,300
Serian Division	**6**	**0**	**104,500**
Sri Aman Division	**0**	**3**	**109,800**
Sri Aman	0	3	77,200
Lubok Antu	0	0	32,600
Betong Division	**3**	**2**	**126,000**
Betong	2	1	72,400
Saratok	1	1	53,600
Sarikei Division	**1**	**0**	**137,400**
Sarikei	1	0	66,500
Meradong	0	0	34,300
Julau	0	0	18,400
Pakan	0	0	18,200
Sibu Division	**11**	**1**	**345,400**
Kanowit	0	0	33,800
Sibu	11	1	284,600
Selangau	0	0	27,000
Mukah Division	0	0	132,700
Daro	0	0	37,300
Dalat	0	0	22,900
Mukah	0	0	51,500
Matu	0	0	21,000
Bintulu Division	**7**	**5**	**262,700**

Bintulu	7	5	226,300
Tatau	0	0	36,400
Kapit Division	**0**	**0**	**132,800**
Belaga	0	0	43,900
Kapit	0	0	64,800
Song	0	0	24,100
Miri Division	**24**	**6**	**428,100**
Miri	24	6	352,300
Marudi	0	0	75,800
Limbang Division	**27**	**4**	**101,600**
Limbang	11	2	56,100
Lawas	16	2	45,500
Total	170	74	2,868,700
Limbang Division	**27**	**4**	**101,600**
Limbang	11	2	56,100
Lawas	16	2	45,500
Total	170	74	2,868,700

14.2 Patient Demographics

There are 244 patients are registered in the MTR in Sarawak up to November 2019 (excluding deceased patients). Fourteen of these patients have received stem cell transplant. Most patients (63.93%) receive treatment at Hospital Umum Sarawak. Tables 14.2 and 14.3, and Figure 14.1 shows the distribution of thalassaemia patients in Sarawak.

There are 20 new cases registered in the MTR in 2019. The new cases are comprised of 10 paediatric patients (1 β-thalassaemia major, 6 HbE/β-thalassaemia, and 3 HbH disease patients) and 10 adult patients.

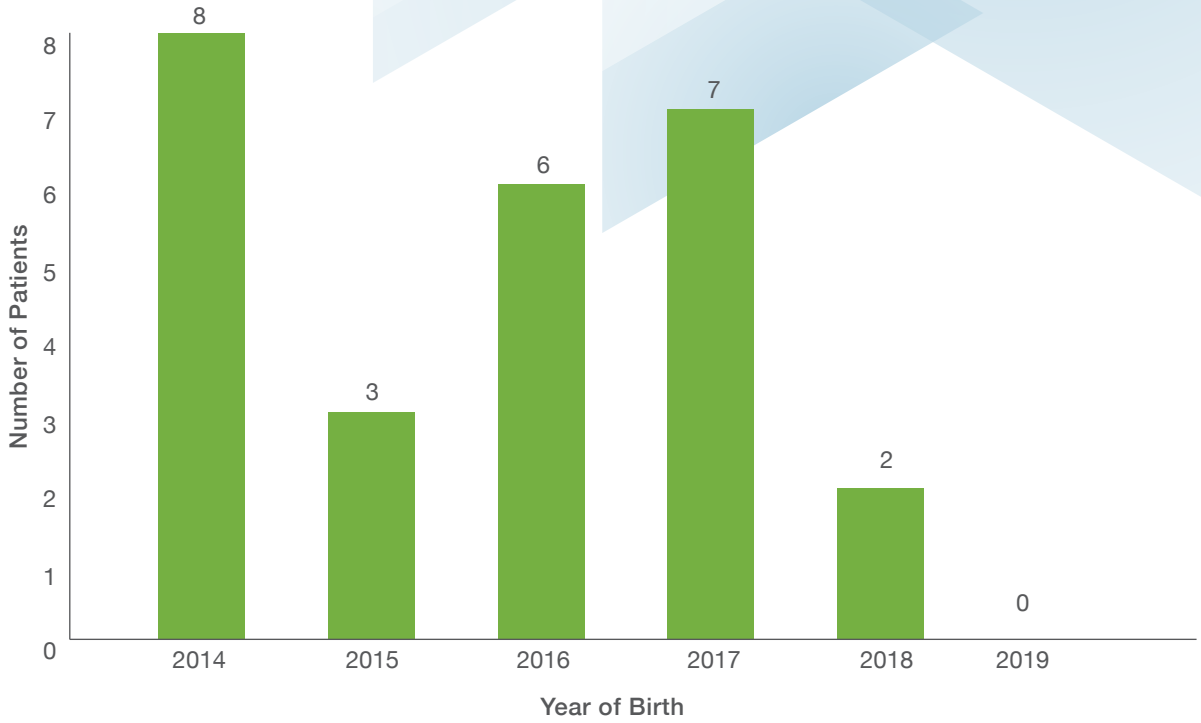
Table 14.2: Distribution of Patients in Sarawak by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Umum Sarawak	156	63.93
Hospital Lundu	2	0.82
Hospital Sarikei	1	0.41
Hospital Sibu	12	4.92
Hospital Miri	30	12.30
Hospital Limbang	13	5.33
Hospital Lawas	18	7.38
Hospital Bintulu	12	4.92
Total	244	100.00

Table 14.3: Distribution of Patients in Sarawak by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	213
Cured by Stem Cell Therapy	14 (2 patients in 2019)
Total	**227**
Lost to Follow-up	17
Total	**244**
Deaths in 2019	0
Cumulative Reported Deaths	21

Figure 14.1: New Thalassaemia Births in Sarawak by Year of Birth



14.2.1 Age Group

The youngest patient in Sarawak is 11 months old (diagnosed with β-thalassaemia major at 9 months of age) and the eldest is 76 years old (diagnosed with HbH disease). Patients below 2 years old are usually transfusion-dependent, even HbE/β-thalassaemia patients. Most of the older patients in Sarawak have HbH disease. They do not require any transfusions and some would require chelation at a later age when the serum ferritin level reaches above 800 ng/mL.

Figure 14.2: Distribution of Patients in Sarawak According to Thalassaemia Type by Age Group

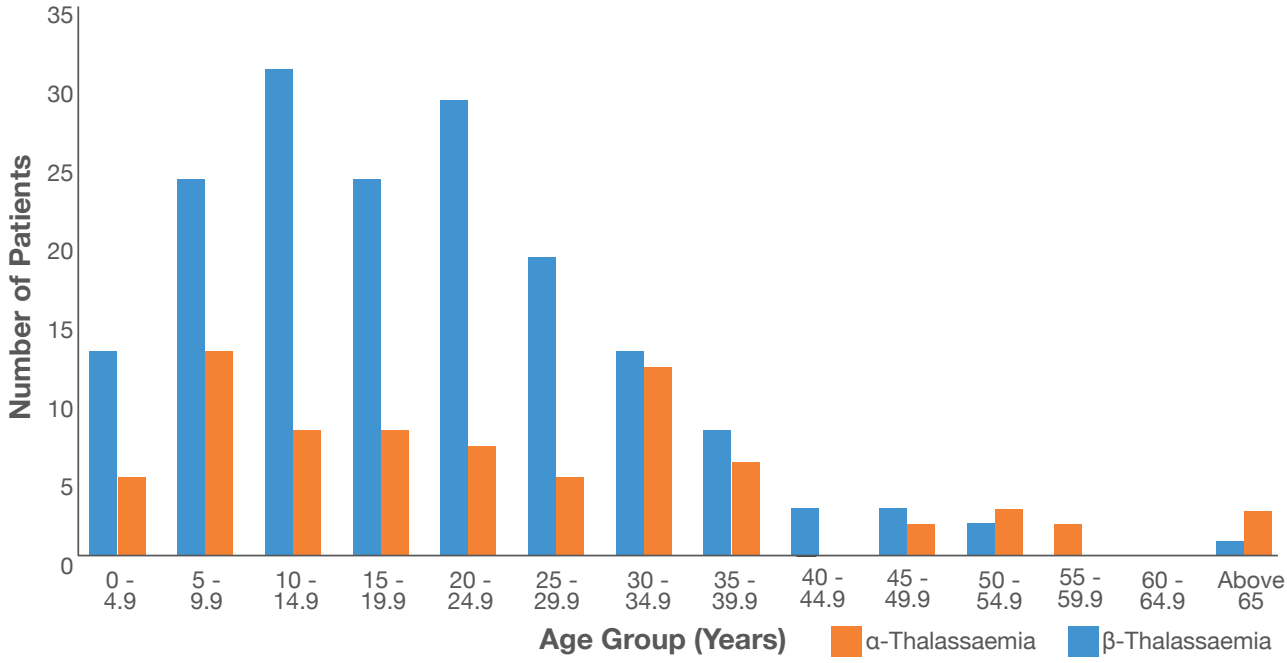
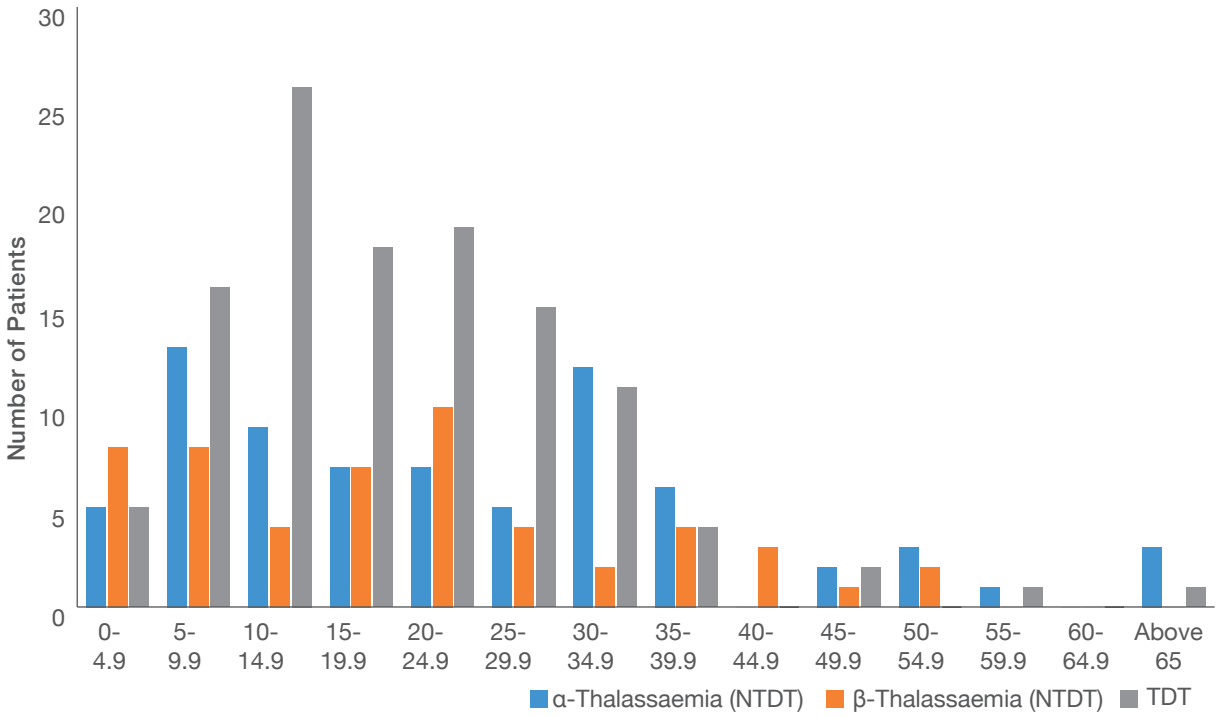


Table 14.4: Distribution of TDT and NTDT (α- and β-Thalassaemia) Patients in Sarawak by Age Group

Age Group (Years)	Number of Patients (TDT)	Number of Patients (NTDT)		Total
		α-Thalassaemia	β-Thalassaemia	
0 – 4.9	5	5	8	18
5 – 9.9	16	13	8	37
10 – 14.9	26	9	4	39
15 – 19.9	18	7	7	32
20 – 24.9	19	7	10	36
25 – 29.9	15	5	4	24
30 – 34.9	11	12	2	25
35 – 39.9	4	6	4	14
40 – 44.9	0	0	3	3
45 – 49.9	2	2	1	5
50 – 54.9	0	3	2	5
55 – 59.9	1	1	0	2
60 – 64.9	0	0	0	0
Above 65	1	3	0	4
Total	118	73	53	244

Figure 14.3: Distribution of TDT and NTDT (α and β-Thalassaemia) Patients in Sarawak by Age Group



The median age group of both TDT and NTDT patients in Sarawak is 15-19.9 years old. The oldest TDT patient is 38 years old. Patients aged 40 years and above are NTDT patients.

Table 14.5: Distribution of Patients in Sarawak According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	94	β-Thalassaemia Major	38	40.43
		β-Thalassaemia Intermedia	4	4.26
		HbE/β-Thalassaemia	26	27.66
		HbH Disease	26	27.66
		Others	0	0.00
15 - 29.9	92	β-Thalassaemia Major	48	52.17
		β-Thalassaemia Intermedia	7	7.61
		HbE/β-Thalassaemia	17	18.48
		HbH Disease	20	21.74
		Others	0	0.00
30 - 44.9	42	β-Thalassaemia Major	15	35.71
		β-Thalassaemia Intermedia	3	7.14
		HbE/β-Thalassaemia	6	14.29
		HbH Disease	18	42.86
		Others	0	0.00
45 - 59.9	12	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	16.67
		HbE/β-Thalassaemia	3	25.00
		HbH Disease	7	58.33
		Others	0	0.00
60 and above	4	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	25.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	3	75.00
		Others	0	0.00
Total			244	

14.2.2 Gender

The gender distribution of thalassaemia patients in Sarawak consists of 125 males (51.23%) and 119 females (48.77%).

Table 14.6: Distribution of Patients in Sarawak According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Umum Sarawak	72	29.51	84	34.43
Hospital Lundu	2	0.82	0	0.00
Hospital Sarikei	1	0.41	0	0.00
Hospital Sibu	4	1.64	8	3.28
Hospital Miri	19	7.79	11	4.51
Hospital Limbang	10	4.10	3	1.23
Hospital Lawas	11	4.51	7	2.87
Hospital Bintulu	6	2.46	6	2.46
Total	125	51.23	119	48.77

14.2.3 Ethnic Group

The ethnic groups with the largest number of thalassaemia patients in Sarawak are Malay (43.45%) and Chinese (36.89%). It must be noted that the Iban patients do not carry the β-globin gene mutation, but instead carry the β-globin gene mutation. Majority of the Kedayan patients carry the Filipino β°-deletion mutation.

Figure 14.4: Distribution of Patients with α-Thalassaemia in Sarawak by Ethnic Group

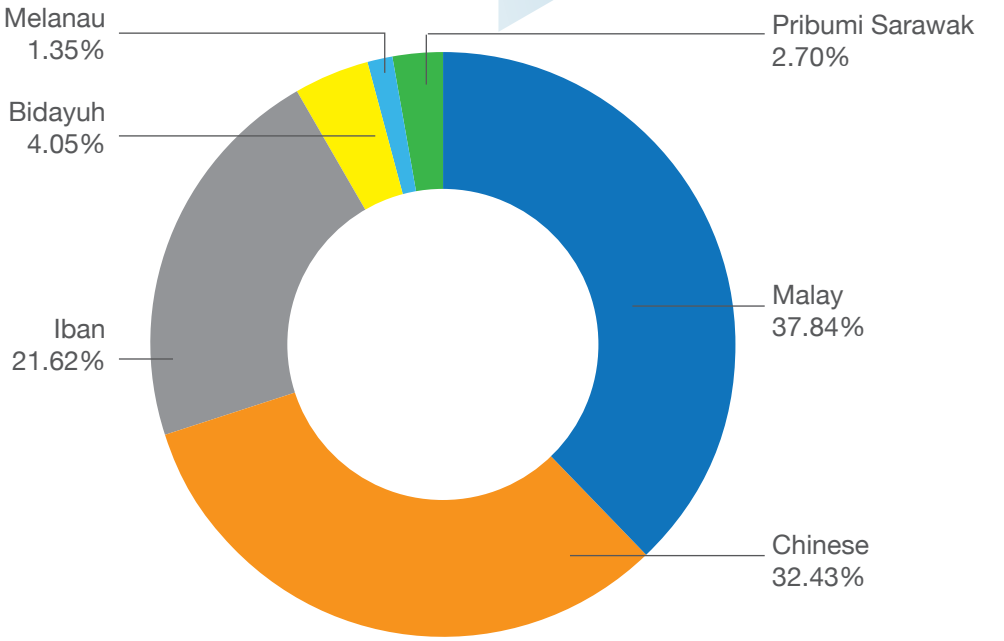


Figure 14.5: Distribution of Patients with β-Thalassaemia in Sarawak by Ethnic Group

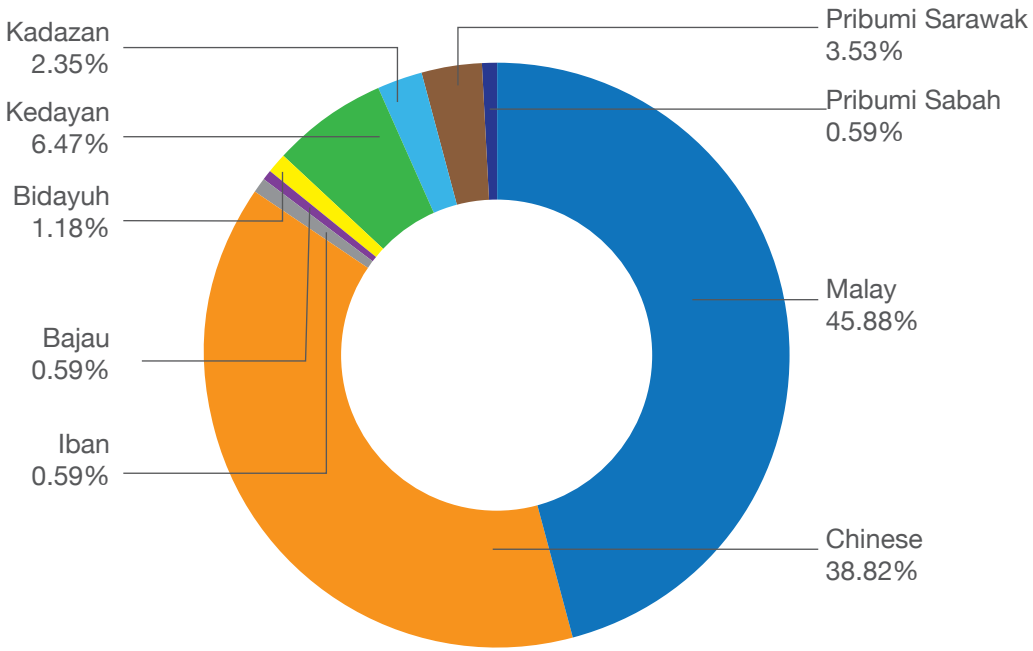


Table 14.7: Distribution of Patients in Sarawak According to Ethnic Group by Centre

Centre	Malay		Chinese		Iban		Bidayuh		Bajau		Kedayan		Kadazan		Melanau	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
HUS Kuching	74	30.33	65	26.64	9	3.69	4	1.64	1	0.41	0	0.00	1	0.41	0	0.00
Hospital Lundu	2	0.82	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Sarikei	0	0.00	0	0.00	1	0.41	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Sibu	0	0.00	12	4.92	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Miri	6	2.46	9	3.69	2	0.82	1	0.41	0	0.00	7	2.87	2	0.82	0	0.00
Hospital Limbang	9	3.69	0	0.00	2	0.82	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Lawas	14	5.74	0	0.00	0	0.00	0	0.00	0	0.00	2	0.82	0	0.00	0	0.00
Hospital Bintulu	1	0.41	4	1.64	3	1.23	0	0.00	0	0.00	2	0.82	1	0.41	1	0.41
Total	106	43.44	90	36.89	17	6.97	5	2.05	1	0.41	11	4.51	4	1.64	1	0.41

14.3 Diagnosis

The main diagnosis of thalassaemia patients in Sarawak is β-thalassaemia major, which make up of 41.39% of the patient population. This is followed by HbH disease (30.33%), HbE/β-thalassaemia (21.31%) and β-thalassaemia intermedia (6.97%). The prevalence of HbH disease is not accurately reflected in this data, as no screening was performed and patients were incidentally noted to be pale. Most HbH patients are asymptomatic.

No β-globin gene mutation was detected among the Iban population. Instead, the Iban harbours the α-globin gene mutations. The only Iban patient reported with β-thalassaemia was in fact adopted, and not of Iban ancestry. The Kedayan, who populates the northern region of Sarawak, carries the Filipino β°-deletion mutation.

Figure 14.6: Distribution of Patients in Sarawak by Diagnosis

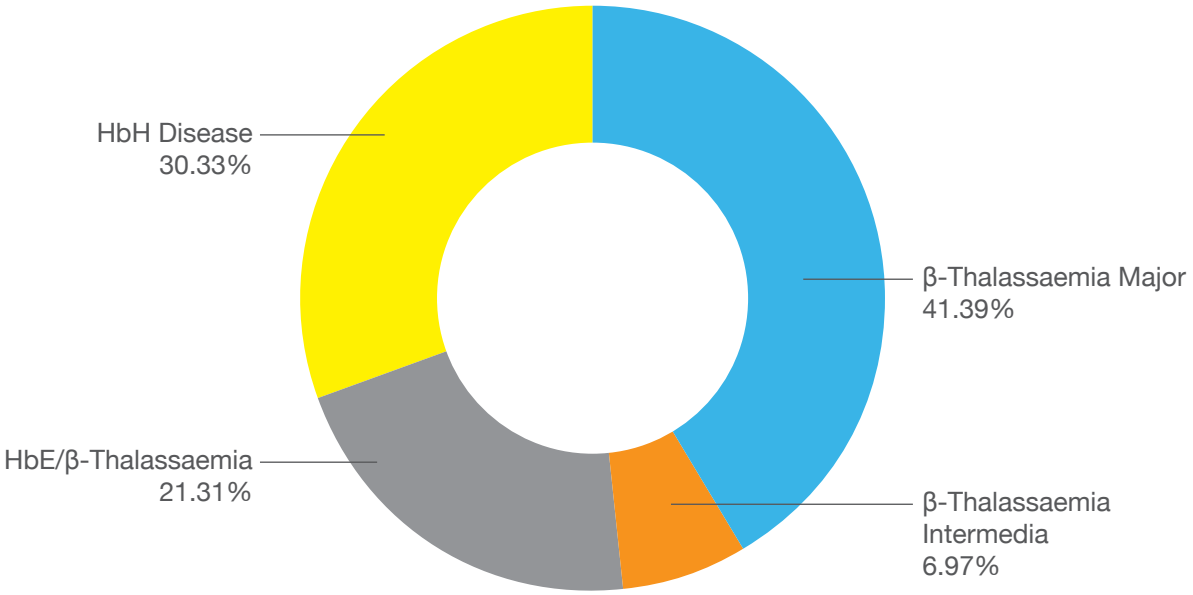


Table 14.8: Distribution of Patients in Sarawak According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	101	Malay	29	11.89
		Chinese	52	21.31
		Iban	0	0.00
		Bidayuh	1	0.41
		Kedayan	10	4.10
		Pribumi Sarawak	5	2.05
		Kadazan-Dusun	4	1.64
β-Thalassaemia Intermedia	17	Malay	7	2.87
		Chinese	7	2.87
		Iban	0	0.00
		Bidayuh	0	0.00
		Kedayan	1	0.41
		Pribumi Sarawak	1	0.41
		Pribumi Sabah	1	0.41
		Kadazan-Dusun	0	0.00
HbE/ β-Thalassaemia	52	Malay	42	17.21
		Chinese	7	2.87
		Iban	1	0.41
		Bajau	1	0.41
		Bidayuh	1	0.41
HbH Disease	74	Malay	28	11.48
		Chinese	24	9.84
		Iban	16	6.56
		Bidayuh	3	1.23
		Melanau	1	0.41
		Pribumi Sarawak	2	0.82
Total			244	100.00

14.4 Treatment

14.4.1 Iron Chelation Therapy

There are 122 patients in Sarawak who are receiving iron chelation therapy. Of these, 43.44% patients are on DFX. All young, newly diagnosed patients in Sarawak are started on DFX monotherapy by 2 years of age if the serum ferritin level is higher than 1000 ng/mL. These patients are also frequently transfused. The patients will only be switched to other drugs if there were side effects or if the patients could not tolerate the drug.

Approximately 27.05% of the patients are on a combination of DFO and DFP, and 17.21% are on DFP monotherapy. These patients are usually older patients, or those who were initially on a combination therapy and were switched to monotherapy when the serum ferritin level reduces. In addition, 8.20% of the patients are on DFO monotherapy and 3.28% are receiving a combination of DFO and DFX. Only 1 (0.82%) patient was on a combination of DFP and DFX.

Patients who receive combination therapy had poor MRI T2* or high serum ferritin levels, which were not responsive to chelation by monotherapy. Nine NTDT patients in Sarawak required chelation. All of them are given DFP monotherapy due to high serum ferritin levels (more than 800 ng/mL). Five of these patients required transfusions every 2-3 months. All 9 patients are aged above 20 years old.

Table 14.9: Distribution of Patients in Sarawak by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	10	8.20
DFP only	21	17.21
DFX only	53	43.44
DFO + DFP	33	27.05
DFP + DFX	1	0.82
DFO + DFX	4	3.28
DFO + DFP + DFX	0	0.00
Total	122	100.00

Table 14.10: Distribution of Patients in Sarawak According to Type of Iron Chelator Received by Centre

Centre	Total Number Of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Umum Sarawak, Kuching	63	DFO only	7	5.74
		DFP only	14	11.48
		DFX only	20	16.39
		DFO + DFP	18	14.75
		DFP + DFX	1	0.82
		DFO + DFX	3	2.46
		DFO + DFP + DFX	0	0.00
Hospital Lundu	2	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	0.82
		DFP + DFX	0	0.00
		DFO + DFX	1	0.82
		DFO + DFP + DFX	0	0.00
Hospital Sibu	9	DFO only	2	1.64
		DFP only	1	0.82
		DFX only	6	4.92
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Miri	18	DFO only	1	0.82
		DFP only	2	1.64
		DFX only	11	9.02
		DFO + DFP	4	3.28
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Limbang	10	DFO only	0	0.00
		DFP only	2	1.64
		DFX only	6	4.92
		DFO + DFP	2	1.64
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Lawas	13	DFO only	0	0.00
		DFP only	1	0.82
		DFX only	5	4.10
		DFO + DFP	7	5.74
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Bintulu	7	DFO only	0	0.00
		DFP only	1	0.82
		DFX only	5	4.10
		DFO + DFP	1	0.82
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			122	100.00

Table 14.11: Distribution of Patients in Sarawak According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	46	DFO only	2	4.35
		DFP only	2	4.35
		DFX only	40	86.96
		DFO + DFP	2	4.35
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15 - 29.9	51	DFO only	9	17.65
		DFP only	6	11.76
		DFX only	9	17.65
		DFO + DFP	24	47.06
		DFP + DFX	3	5.88
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 44.9	16	DFO only	0	0.00
		DFP only	5	31.25
		DFX only	4	25.00
		DFO + DFP	6	37.50
		DFP + DFX	0	0.00
		DFO + DFX	1	6.25
		DFO + DFP + DFX	0	0.00
45 - 59.9	7	DFO only	0	0.00
		DFP only	6	85.71
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	1	14.29
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

60 and above	2	DFO only	0	0.00
		DFP only	2	100.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
			122	

14.4.2 Serum Ferritin Level

A total of 177 patients in Sarawak had their serum ferritin level recorded in 2019. Of these, 103 (58.19%) are TDT patients and 74 (41.81%) are NTDT patients. Eighty-one (45.76%) patients have a serum ferritin level lower than 1000 ng/mL, 55 patients (31.07%) have a serum ferritin level between 1000-2499 ng/mL, and 24 (13.56%) patients have a serum ferritin level between 2500-4999 ng/mL. Besides that, 14 (7.91%) patients have a serum ferritin level between 5000-9999 ng/mL and 3 (1.69%) patients recorded the highest serum ferritin levels, above 10,000 ng/mL. Thirty-three patients, mostly NTDT, do not have their serum ferritin level recorded.

Most patients (136 out of 177 patients, 76.84%) from regional centres and tertiary centres have a serum ferritin level lower than 2500 ng/mL, whereas 41 out of 177 patients (23.16%) have a serum ferritin level above 2500 ng/mL.

The patients in Hospital Lawas and Hospital Limbang consistently recorded the highest serum ferritin levels. Adherence to chelation therapy is a recurring problem for patients in these regions. Further study is needed to ascertain the causes resulting in poor adherence.

Table 14.12: Distribution of Patients in Sarawak According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Umum Sarawak, Kuching	115	66	37.29	31	17.51	14	7.91	3	1.69	1	0.56
Hospital Miri	22	10	5.65	9	5.08	1	0.56	2	1.13	0	0.00
Hospital Sibu	7	0	0.00	6	3.39	0	0.00	1	0.56	0	0.00
Hospital Bintulu	6	1	0.56	4	2.26	0	0.00	0	0.00	1	0.56
Hospital Lawas	15	2	1.13	3	1.69	2	1.13	8	4.52	0	0.00
Hospital Limbang	11	2	1.13	2	1.13	6	3.39	0	0.00	1	0.56
Hospital Lundu	1	0	0.00	0	0.00	1	0.56	0	0.00	0	0.00
Total	177	81	45.76	55	31.07	24	13.56	14	7.91	3	1.69

Figure 14.7: Distribution of Patients in Sarawak According to Most Recent Serum Ferritin Level

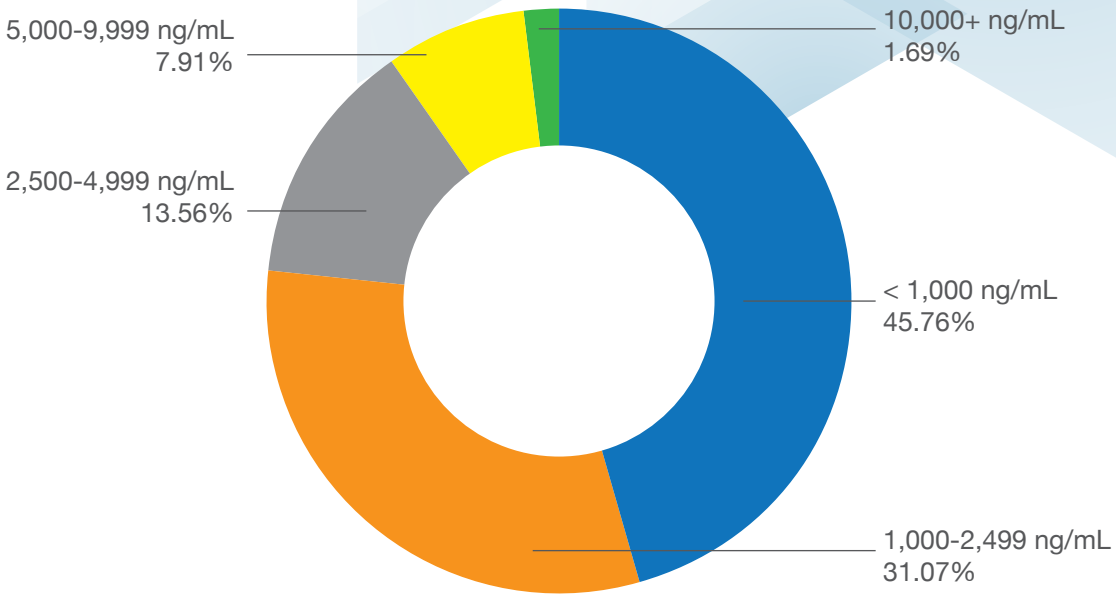


Table 14.13: Distribution of TDT Patients in Sarawak According to Most Recent Serum Ferritin Level by Age Group

Age Group (Years)	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
0 - 4.9	4	1	0.97	2	1.94	1	0.97	0	0.00	0	0.00
5 - 9.9	16	1	0.97	12	11.65	3	2.91	0	0.00	0	0.00
10 - 14.9	25	5	4.85	14	13.59	4	3.88	2	1.94	0	0.00
15 - 19.9	16	3	2.91	7	6.80	4	3.88	2	1.94	0	0.00
20 - 24.9	16	0	0.00	6	5.83	5	4.85	5	4.85	0	0.00
25 - 29.9	13	3	2.91	3	2.91	3	2.91	3	2.91	1	0.97
30 - 34.9	8	3	2.91	3	2.91	0	0.00	1	0.97	1	0.97
35 - 39.9	4	3	2.91	1	0.97	0	0.00	0	0.00	0	0.00
Above 40	1	0	0.00	0	0.00	1	0.97	0	0.00	0	0.00
Total	103	19	18.45	48	46.60	21	20.39	13	12.62	2	1.94

Twenty patients with TDT in Sarawak are below 10 years old. Sixteen of these patients have a serum ferritin level < 2500 ng/mL.

Table 14.14: Distribution of NTDT Patients in Sarawak According to Most Recent Serum Ferritin Level by Age Group

Age Group (Years)	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
0 - 4.9	10	10	13.51	0	0.00	0	0.00	0	0.00	0	0.00
5 - 9.9	14	14	18.92	0	0.00	0	0.00	0	0.00	0	0.00
10 - 14.9	7	6	8.11	1	1.35	0	0.00	0	0.00	0	0.00
15 - 19.9	12	10	13.51	1	1.35	1	1.35	0	0.00	0	0.00
20 - 24.9	5	4	5.41	0	0.00	0	0.00	1	1.35	0	0.00
25 - 29.9	1	1	1.35	0	0.00	0	0.00	0	0.00	0	0.00
30 - 34.9	6	5	6.76	1	1.35	0	0.00	0	0.00	0	0.00
35 - 39.9	5	4	5.41	1	1.35	0	0.00	0	0.00	0	0.00
Above 40	14	8	10.81	3	4.05	2	2.70	0	0.00	1	1.35
Total	74	62	83.78	7	9.46	3	4.05	1	1.35	1	1.35

Majority of NTDT patients in Sarawak have a serum ferritin level below 1000 ng/mL. Only 1 patient (55 years old, HbH disease-Hb Adana) in Sarawak has a serum ferritin level higher than 10,000 ng/mL.

Table 14.15: Distribution of Patients in Sarawak According to Transfusion Status by Centre

Centre	Number of Patients (TDT)	Number of Patients (NTDT)
Hospital Umum Sarawak, Kuching	59	97
Hospital Lundu	2	0
Hospital Sarikei	0	1
Hospital Sibu	9	3
Hospital Miri	18	12
Hospital Limbang	10	3
Hospital Lawas	13	5
Hospital Bintulu	7	5
Total	118	126

14.5 Complications and Deaths

Table 14.16: Infectious Complications in Thalassaemia Patients Above 15 Years Old in Sarawak

Infections	Hepatitis B	2
	Hepatitis C	10
	HIV	0

There are 58 TDT and 50 NTDT patients above 15 years old in Sarawak.

One adult patient was diagnosed with hepatitis B, and another 3 patients were diagnosed with hepatitis C in 2019. It is unknown whether the transmissions were due to previous transfusions, or due to high-risk behaviours. The introduction of NAT testing among donors since January 2019 is expected to control the number of infective complications related to blood products transfusion.

Twenty-seven out of 108 patients (25%) were diagnosed with delayed puberty and were put on hormonal replacement therapy. These patients have been poorly chelated in the past. Fifty four patients (50%) were noted to have short stature, but further information is required to determine whether this is secondary to growth hormone deficiency or familial short stature. It is expected that with better chelation, the number of patients with the endocrinological complications will reduce.

Of the 19 patients who underwent dual-energy X-ray absorptiometry (DEXA) scan, 84.21% showed evidence of either osteopenia or osteoporosis. All patients with delayed puberty have osteopenia and osteoporosis.

Eighty patients had T2* performed between 2016 and October 2019, and 81.25% of these patients have a normal T2*. Some of these patients had shown an initial T2* of less than 20 ms, but has since improved over the years with intensive chelation. In 2019, 11 patients had T2* scans performed up to October, and another 11 patients had their T2* scans performed in November and December 2019. Two of these patients had T2* less than 10 ms. These 2 patients were known to be poorly compliant to chelation therapy for many years. An additional 2 patients had a T2* of between 10-20 ms. Most of these patients have endocrine problems.

Table 14.17: Endocrine Complications in Patients Above 15 Years Old Receiving Iron Chelation Therapy in Sarawak

Endocrine	Delayed Puberty	27 (3 post BMT)
	DM	6 (2 died, 1 post BMT)
	Hypothyroidism	7
	Growth Retardation	54
Bone Health (DEXA Scan)	Osteoporosis	16
	Normal DEXA scan	3

Table 14.18: Cardiac MRI T2* Results in Sarawak

Cardiac (MRI T2*)	< 10 ms	6
	10–20 ms	9
	> 20 ms	65

Eighty patients had MRI T2* performed between 2016 and October 2019.

Table 14.19: Cumulative Causes of Death in Sarawak

Causes of Death	Number of Patients	Percentage (%)
Infections	4	19.05
Cardiac Complications	14	66.67
Thalassaemia (Anaemia/Alloimmunisation)	1	4.76
Others	1 (died at age 88)	4.76
Liver Disease	1	4.76
Total	21	100.00

A total of 21 patients had died in Sarawak since 1997. The figure remained unchanged since 2018, as there were no deaths recorded in 2019.

14.6 Conclusion

Based on the data collection, there was a 9.4% increment in the number of new cases registered in the MTR in 2019. Of the 20 new cases registered, 9 are children (3 diagnosed with HbH disease, 1 with β-thalassaemia major, and 5 with HbE/β-thalassaemia). Most of these new patients have a family member with known thalassaemia. Therefore, genetic counselling of patients must be improved. Interracial marriage also contributes to new thalassaemia cases. The median age group of patients in Sarawak is 15-19.9 years old, which remained the same as in 2018 as new cases are still being registered in both the paediatric and adult cohorts each year.

Most patients (76.8%) in Sarawak have a serum ferritin level < 2500 ng/mL. In the TDT cohort, 65% of patients have a serum ferritin level < 2500 ng/mL, of which 19.4% have a serum ferritin level < 1000 ng/mL. A majority of patients with an elevated serum ferritin level (> 2500 ng/mL) are adult patients between 20-30 years old, who are in the their most productive years. This observation is correlated with the decline in adherence to iron chelation therapy. Further study is needed to investigate the factors resulting in poor therapeutic compliance, e.g. failure in transition from paediatric to adult care.

There was some improvement in the serum ferritin level of the patients from Limbang following the placement of a paediatrician in Hospital Limbang beginning 2019. Patient care was better coordinated with the presence of medical staff experienced in managing thalassaemia patients. However, there still remains a group of patients in this region who are non-compliant to treatment. It is important to delve into the causes of non-compliance, e.g. patients' personal/cultural beliefs regarding treatment, lack of dedicated pharmacists to counsel on adherence, geographical difficulties to access healthcare facilities, etc. On the other hand, Lawas consistently recorded the highest number of patients with serum ferritin level > 2500 mg/mL. It is likely that the patients in Lawas experience the same challenges faced by the patients in Limbang. Paediatricians/physicians in Hospital Limbang or Miri may consider taking over the care of thalassaemia patients in Lawas in the future.

A significant number of patients were not well-chelated in their youth; therefore, a substantial number of these patients experience endocrine complications, especially affecting growth and development. Fifty-eight of TDT patients are above 15 years old. Of these, approximately 50% have delayed puberty and short stature. Bone health also remains a major problem. All patients with delayed puberty suffered from osteoporosis or osteopenia. We, however, could not offer DEXA scan to all patients, since the sole DEXA machine in the state hospital has been in disrepair for several years. Better chelation during childhood is expected to reduce endocrine complications over the years.

MRI T2* scans were attempted on all chelated patients above 10 years of age. Of the 80 patients who had a T2* scan performed, 18.75% have an MRI T2* result below 20 ms. Some patients had an MRI T2* result of < 20 ms, but was reversed over the years with intensive chelation. Since the beginning of the registry, Sarawak has recorded 21 deaths, mostly due to cardiac complications. Of note, there was a mortality attributed to severe haemolytic anaemia due to alloimmunisation. There is a need to examine the frequency of, and factors leading to, alloimmunisation, especially in Limbang and Lawas where alloimmunisation is more prevalent. There were no deaths recorded in 2019.

15 Selangor

15.1 Introduction

Selangor is the eighth largest state in Peninsular Malaysia with an estimated population of 6,530,000 in 2019 (Department of Statistics Malaysia). Selangor is divided into 9 administrative districts, which are Gombak, Hulu Langat, Hulu Selangor, Klang, Kuala Langat, Kuala Selangor, Petaling, Sabak Bernam and Sepang.

There are 1249 living thalassaemia patients in Selangor. Most thalassaemia patients in the state receive transfusions and follow-up care in the hospitals listed in Table 15.1. Most hospitals carry out transfusions in their Ambulatory Care Centre. Hospitals without an Ambulatory Care Centre carry out transfusions in the wards.

15.2 Patient Demographics

There are 1331 thalassaemia patients recorded in Selangor. These patients receive treatment at the 11 centres listed in Table 15.1. Hospital Ampang handles the majority of thalassaemia patients in Selangor, as all paediatric patients from the Institute of Paediatrics, Hospital Kuala Lumpur (IPHKL) and Hospital Selayang are transferred to this centre when they turn 18 years old. Hospital Tengku Ampuan Rahimah (HTAR), Klang is the centre of referral for the Selangor Central Zone hospital cluster and has the second highest number of patients in Selangor.

Table 15.1: Distribution of Patients in Selangor by Centre

Centre	Number of Living Patients	%
Hospital Ampang	731	58.53
Hospital Banting	19	1.52
Hospital Kajang	46	3.68
Hospital Kuala Kubu Bharu	3	0.24
Hospital Selayang	94	7.53
Hospital Serdang	43	3.44
Hospital Shah Alam	17	1.36
Hospital Sungai Buloh	60	4.80
Hospital Tanjung Karang	3	0.24
Hospital Tengku Ampuan Jemaah, Sabak Bernam	5	0.40
Hospital Tengku Ampuan Rahimah, Klang	228	18.25
Total	1249	100.00

Table 15.2: Distribution of Patients in Selangor by Vital Status

Vital Status	Number of Patients
Alive and On Active Treatment	1093
Haemopoietic Stem Cell Transplant	10
Total	**1103**
Lost to Follow-up	146
Total	**1249**
Deaths in 2019	14
Cumulative Reported Deaths	82

Table 15.3: Cumulative Causes of Death in Selangor Since 2007

Causes of Death	Number of Patients	Percentage (%)
Infections	27	34.18
Cardiac Causes	30	37.97
Liver Disease	8	10.13
Tumours	4	5.06
Endocrine Complication	1	1.27
Motor Vehicle Accident (MVA)	5	6.33
Arrhythmias	1	1.27
Renal Complications	1	1.27
Thalassaemia	2	2.53
Total	79	100.00

Three patients have an unknown cause of death due to unavailable data.

15.2.1 Age Group

Approximately 35.23% of thalassaemia patients in Selangor are below 20 years old, and fall under the paediatric and adolescent age categories. Adult patients above 21 years old form 64.77% of the thalassaemia cases in Selangor. The oldest patient in Selangor is an 87-year-old diagnosed with β-thalassaemia intermedia. Meanwhile, the youngest patient is a 5-month-old Malay male diagnosed with β-thalassaemia major, and is followed up in the HTAR, Klang.

Figure 15.1: Distribution of Patients in Selangor by Age Group

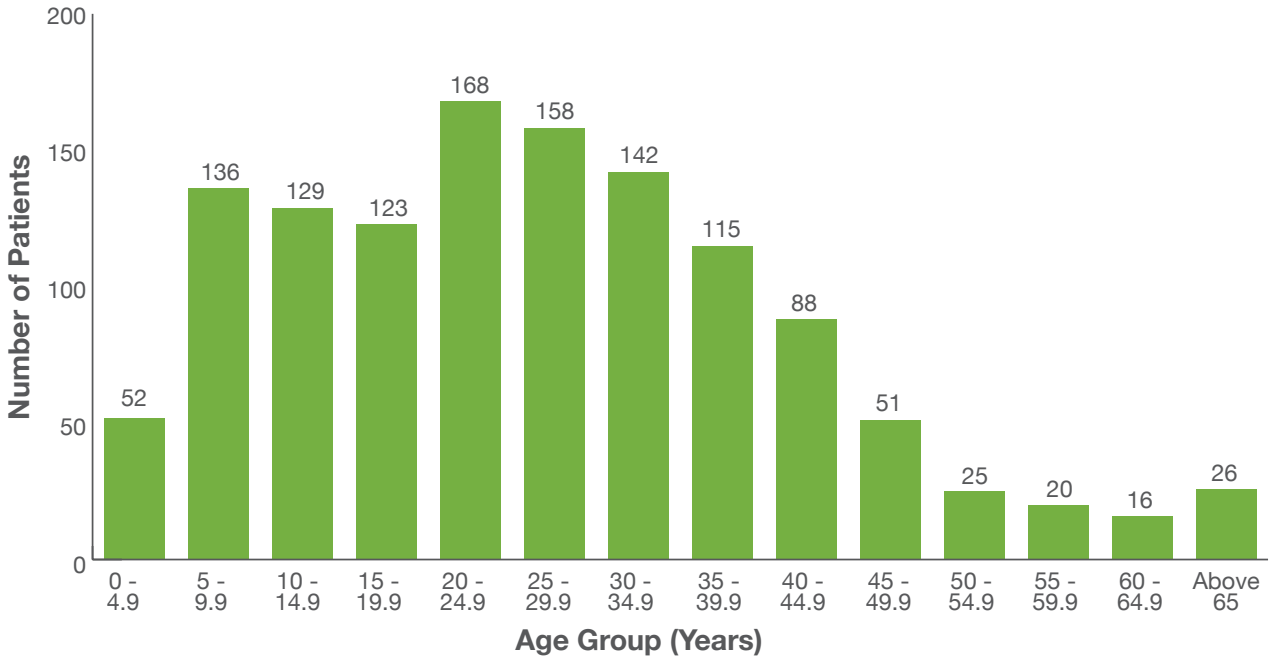


Table 15.4: Distribution of Patients in Selangor According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	317	β-Thalassaemia Major	60	18.93
		β-Thalassaemia Intermedia	154	48.58
		HbE/β-Thalassaemia	19	5.99
		HbH Disease	59	18.61
		Others	25	7.89
15 - 29.9	449	β-Thalassaemia Major	108	24.05
		β-Thalassaemia Intermedia	202	44.99
		HbE/β-Thalassaemia	31	6.90
		HbH Disease	97	21.60
		Others	11	2.45
30 - 44.9	345	β-Thalassaemia Major	51	14.78
		β-Thalassaemia Intermedia	147	42.61
		HbE/β-Thalassaemia	22	6.38
		HbH Disease	111	32.17
		Others	14	4.06
45 - 59.9	96	β-Thalassaemia Major	3	3.13
		β-Thalassaemia Intermedia	34	35.42
		HbE/β-Thalassaemia	15	15.63
		HbH Disease	40	41.67
		Others	4	4.17
60 and above	42	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	10	23.81
		HbE/β-Thalassaemia	5	11.90
		HbH Disease	26	61.90
		Others	1	2.38
Total			1249	

15.2.2 Gender

There are more female than male thalassaemia patients in Selangor, at 674 (53.96%) female and 575 (46.04%) male patients, respectively.

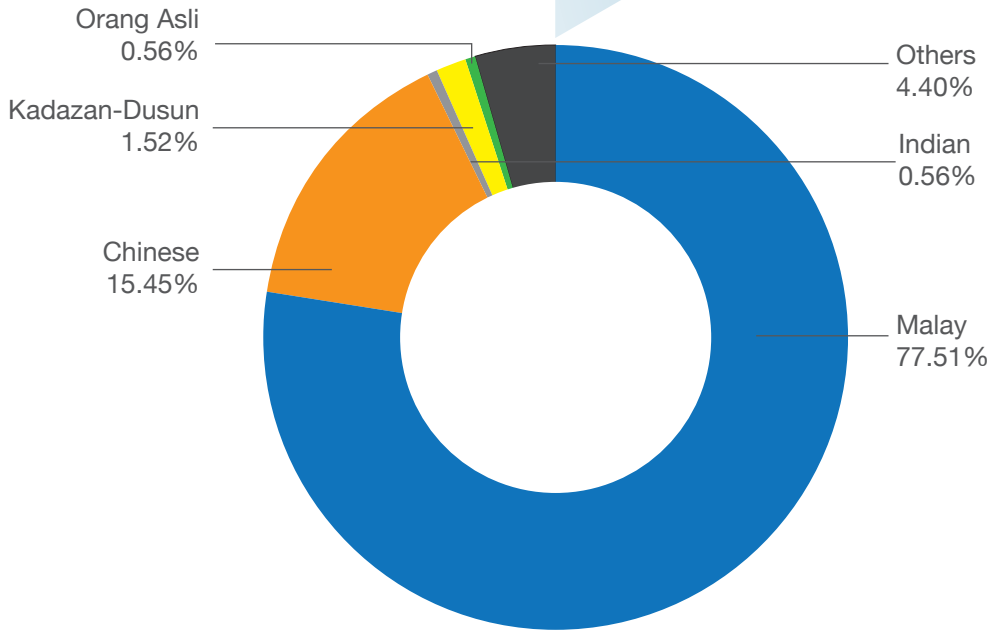
Table 15.5: Distribution of Patients in Selangor According to Gender by Centre

Centre	Male		Female	
	Number	%	Number	%
Hospital Ampang	295	23.62	436	34.91
Hospital Banting	15	1.20	4	0.32
Hospital Kajang	26	2.08	20	1.60
Hospital Kuala Kubu Bharu	2	0.16	1	0.08
Hospital Selayang	49	3.92	45	3.60
Hospital Serdang	25	2.00	18	1.44
Hospital Sungai Buloh	39	3.12	21	1.68
Hospital Tanjung Karang	0	0.00	3	0.24
Hospital Tengku Ampuan Jemaah, Sabak Bernam	3	0.24	2	0.16
Hospital Tengku Ampuan Rahimah, Klang	111	8.89	117	9.37
Hospital Shah Alam	10	0.80	7	0.56
Total	575	46.04	674	53.96

15.2.3 Ethnic Group

Malay patients form the largest group of patients in Selangor with 968 patients (77.51%), followed by the Chinese (193 patients, 15.45%), Indian (7 patients, 0.56%), Kadazan-Dusun (19 patients, 1.52 %), and Orang Asli (7 patients, 0.56%). Other ethnic groups (foreigners, Murut, Melanau and Thai) contributes 55 patients (4.40%) in Selangor.

Figure 15.2: Distribution of Patients in Selangor by Ethnic Group



15.3 Diagnosis

Based on Figure 15.3, HbE/β-thalassaemia has the highest number of patients in Selangor (547 patients, 43.80%). This is followed by HbH disease (333 patients, 26.66%), β-thalassaemia major (222 patients, 17.77%), and β-thalassaemia intermedia (92 patients, 7.37%). The remaining 55 patients (4.40%) are being followed up for other forms of haemoglobinopathy. Based on Table 15.6, HbE/β-thalassaemia is the predominant diagnosis among Malay patients, whereas HbH disease is predominant among the Chinese.

Figure 15.3: Distribution of Patients in Selangor by Diagnosis

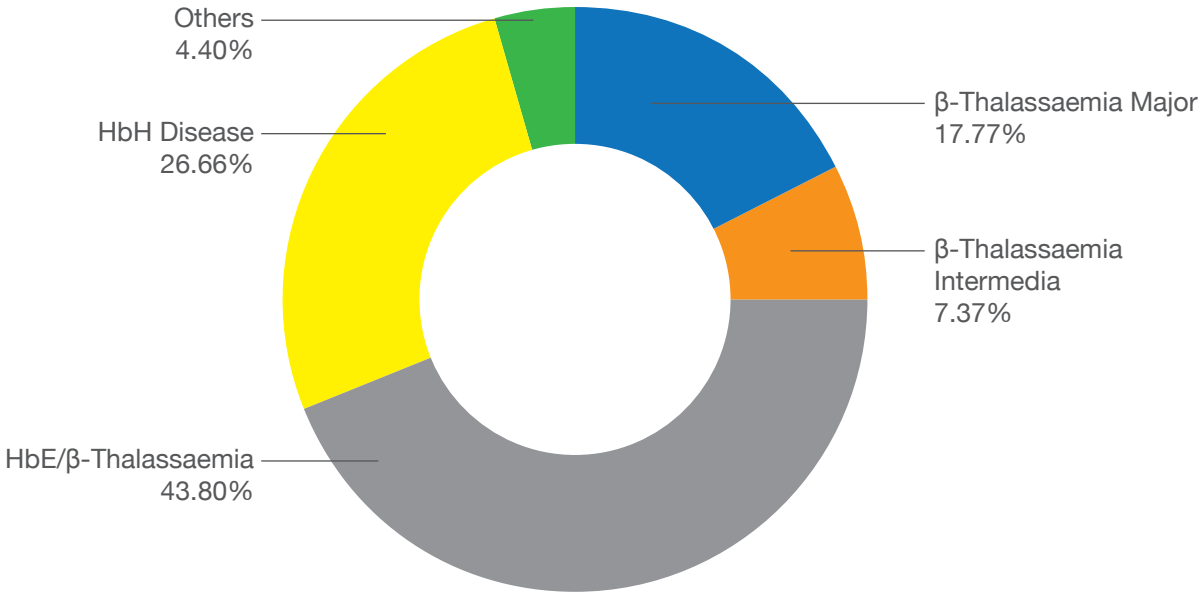


Table 15.6: Distribution of Patients in Selangor According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	222	Malay	122	9.77
		Chinese	69	5.52
		Indian	2	0.16
		Kadazan-Dusun	14	1.12
		Others	15	1.20
β-Thalassaemia Intermedia	92	Malay	73	5.84
		Chinese	12	0.96
		Indian	1	0.08
		Kadazan-Dusun	1	0.08
		Others	5	0.40
HbE/ β-Thalassaemia	547	Malay	496	39.71
		Chinese	20	1.60
		Indian	1	0.08
		Kadazan-Dusun	2	0.16
		Orang Asli	3	0.24
		Others	25	2.00
HbH Disease	333	Malay	237	18.98
		Chinese	84	6.73
		Indian	1	0.08
		Kadazan-Dusun	1	0.08
		Orang Asli	4	0.32
		Others	6	0.48
Others	55	Malay	40	3.20
		Chinese	8	0.64
		Indian	2	0.16
		Kadazan-Dusun	2	0.16
		Orang Asli	0	0.00
		Others	3	0.24
Total	1249		1249	100.00

15.4 Treatment

15.4.1 Iron Chelation Therapy

Once iron overload is identified by evaluating serum ferritin level and MRI T2* results, iron chelation therapy is initiated to remove the iron. According to Table 15.7, 809 patients in Selangor receive iron chelation therapy. Of these, 328 patients (40.54%) are on oral DFP monotherapy, 169 patients (20.89%) are on DFX monotherapy, and 74 patients (9.15%) patients are on DFO monotherapy. A total of 208 patients (25.71%) are on DFO + DFP combination therapy, 22 patients (2.72%) are on DFO + DFX combination therapy, and 7 patients (0.87%) are on DFP + DFX combination therapy. Only 1 patient (0.12%) receives all three types of chelators (DFO + DFP + DFX).

Table 15.7: Distribution of Patients in Selangor by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	74	9.15
DFP only	328	40.54
DFX only	169	20.89
DFO + DFP	208	25.71
DFP + DFX	7	0.87
DFO + DFX	22	2.72
DFO + DFP + DFX	1	0.12
Total	809	100.00

As shown in Table 15.8, DFX monotherapy is the most common iron chelator used in Hospital Klang, Hospital Serdang, Hospital Selayang and Hospital Shah Alam, as thalassaemia patients in these hospitals are of the paediatrics age group. On the other hand, Hospital Ampang utilises DFP monotherapy most frequently, as they mainly treat adult patients. Most patients who had been treated with DFX as a paediatric patient are usually switched to DFO/DFP upon transfer to adult care, as there is no additional funding to support DFX in this cohort of patients.

Table 15.8: Distribution of Patients in Selangor According to Type of Iron Chelator Received by Centre

Centre	Total Number Of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Ampang	487	DFO only	34	4.20
		DFP only	264	32.63
		DFX only	22	2.72
		DFO + DFP	152	18.79
		DFP + DFX	8	0.99
		DFO + DFX	6	0.74
		DFO + DFP + DFX	1	0.12
Hospital Banting	12	DFO only	4	0.49
		DFP only	3	0.37
		DFX only	3	0.37
		DFO + DFP	2	0.25
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kajang	23	DFO only	7	0.87
		DFP only	1	0.12
		DFX only	5	0.62
		DFO + DFP	9	1.11
		DFP + DFX	0	0.00
		DFO + DFX	1	0.12
		DFO + DFP + DFX	0	0.00
Hospital Kuala Kubu Bharu	3	DFO only	1	0.12
		DFP only	1	0.12
		DFX only	0	0.00
		DFO + DFP	1	0.12
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Selayang	35	DFO only	3	0.37
		DFP only	3	0.37
		DFX only	25	3.09
		DFO + DFP	1	0.12
		DFP + DFX	0	0.00
		DFO + DFX	3	0.37
		DFO + DFP + DFX	0	0.00
Hospital Serdang	29	DFO only	9	1.11
		DFP only	0	0.00
		DFX only	20	2.47
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Shah Alam	9	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	9	1.11
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Sungai Buloh	44	DFO only	2	0.25
		DFP only	13	1.61
		DFX only	7	0.87
		DFO + DFP	21	2.60
		DFP + DFX	0	0.00
		DFO + DFX	1	0.12
		DFO + DFP + DFX	0	0.00
Hospital Tanjung Karang	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tengku Ampuan Jemaah, Sabak Bernam	3	DFO only	2	0.25
		DFP only	0	0.00
		DFX only	1	0.12
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Tengku Ampuan Rahimah, Klang	164	DFO only	12	1.48
		DFP only	43	5.32
		DFX only	77	9.52
		DFO + DFP	22	2.72
		DFP + DFX	1	0.12
		DFO + DFX	9	1.11
		DFO + DFP + DFX	0	0.00
Total			809	100.00

The type of iron chelator received by patients according to age group is shown in Table 15.9. For patients aged 20 years old and above, DFP is the most common iron chelator used, sometimes in combination with DFO.

Table 15.9: Distribution of Patients in Selangor According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0–14.9	193	DFO only	20	10.36
		DFP only	22	11.40
		DFX only	121	62.69
		DFO + DFP	22	11.40
		DFP + DFX	2	1.04
		DFO + DFX	6	3.11
		DFO + DFP + DFX	0	0.00
15–29.9	312	DFO only	30	9.62
		DFP only	115	36.86
		DFX only	35	11.22
		DFO + DFP	116	37.18
		DFP + DFX	11	3.53
		DFO + DFX	4	1.28
		DFO + DFP + DFX	1	0.32
30–44.9	224	DFO only	19	8.48
		DFP only	137	61.16
		DFX only	4	1.79
		DFO + DFP	59	26.34
		DFP + DFX	1	0.45
		DFO + DFX	4	1.79
		DFO + DFP + DFX	0	0.00
45–59.9	59	DFO only	5	8.47
		DFP only	38	64.41
		DFX only	5	8.47
		DFO + DFP	10	16.95
		DFP + DFX	0	0.00
		DFO + DFX	1	1.69
		DFO + DFP + DFX	0	0.00
60 and above	21	DFO only	0	0.00
		DFP only	16	76.19
		DFX only	4	19.05
		DFO + DFP	1	4.76
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			809	

15.4.2 Serum Ferritin Level

Serum ferritin level was measured to evaluate iron overload upon regular blood transfusion. A total of 510 TDT patients had their serum ferritin levels measured in 2019. Eighty-four patients (16.47%) have a serum ferritin level of lower than 1000 ng/mL. There are 169 patients (33.14%) with a serum ferritin level between 1000-2499 ng/mL. A total of 155 patients (30.39%) have a serum ferritin level lower than 5000 ng/mL, while 84 patients (16.47%) have a serum ferritin level lower than 10,000 ng/mL. Eighteen patients (3.53%) have a serum ferritin level more than 10,000 ng/mL. Most of the patients with high serum ferritin levels are adolescents and adults, who are likely to be non-compliant to iron chelating agents.

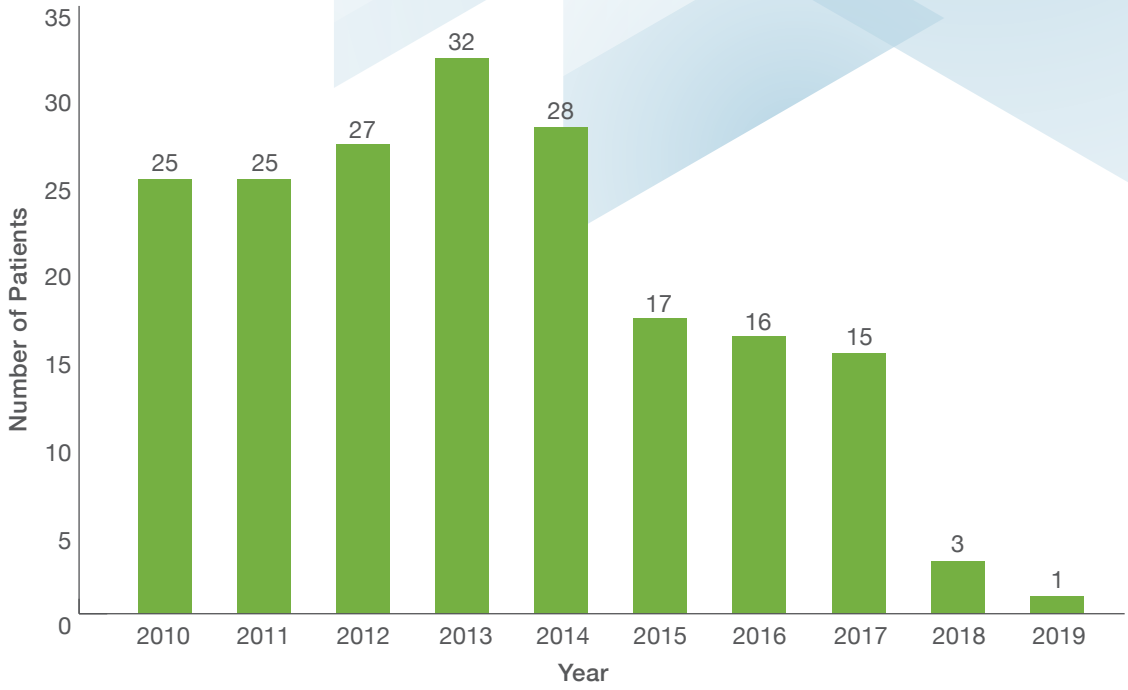
Table 15.10: Distribution of Patients in Selangor According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Ampang	265	48	9.41	82	16.08	61	11.96	60	11.76	14	2.75
Hospital Banting	16	4	0.78	8	1.57	3	0.59	1	0.20	0	0.00
Hospital Kajang	11	0	0.00	3	0.59	5	0.98	2	0.39	1	0.20
Hospital Kuala Kubu Baru	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Selayang	29	5	0.98	10	1.96	13	2.55	1	0.20	0	0.00
Hospital Serdang	7	0	0.00	6	1.18	1	0.20	0	0.00	0	0.00
Hospital Shah Alam	8	3	0.59	4	0.78	1	0.20	0	0.00	0	0.00
Hospital Sungai Buloh	42	8	1.57	12	2.35	19	3.73	2	0.39	1	0.20
Hospital Tanjung Karang	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Sabak Bernam	1	0	0.00	1	0.20	0	0.00	0	0.00	0	0.00
HTAR, Klang	131	16	3.14	43	8.43	52	10.20	18	3.53	2	0.39
Total	510	84	16.47	169	33.14	155	30.39	84	16.47	18	3.53

15.4.3 Thalassaemia Births

The number of thalassaemia births in Selangor showed an increase from 2010 to 2013. However, new thalassaemia births have since started reducing in trend, from 32 new births in 2013 to only 1 new patient in 2019.

Figure 15.4: New Thalassaemia Births in Selangor by Year



15.5 Conclusion

In Selangor, male and female genders are equally affected by thalassaemia. Malay patients form the largest number of patients in Selangor. The age group of between 20-24.9 years has the highest number of patients; which is essentially the reproductive age group. The majority of adult patients are treated in Hospital Ampang, as it is the country's referral centre for adult haematology. New thalassaemia births in Selangor have decreased from year 2013 to 2019. We hope that the reducing trend will continue with the implementation of the National Thalassaemia Screening Programme.

Most patients in Selangor are diagnosed with HbE/β-thalassaemia. Other haemoglobinopathies include β-thalassaemia major, HbH disease, β-thalassaemia intermedia and more. All these diagnoses were based on haemoglobin analysis and DNA analysis for identification of specific mutations.

A significant group of patients receiving iron chelation therapy in Selangor are on DFP monotherapy, and the most common combination therapy is DFO/DFP. Reinforcement of compliance to iron chelators is important to prevent iron overload complications, such as cardiac complications.

About half of TDT patients in Selangor have a serum ferritin level of between 1000-2500 ng/mL. Most NTDT patients' serum ferritin levels are not regularly monitored and their appointments are at longer intervals compared to TDT patients. In order to prevent thalassaemia from becoming a disease burden to the country, carriers must be advised to take a more proactive approach in choosing their future partners. Parents, on the other hand, play an important role in ensuring their children choose the appropriate partners. More premarital thalassaemia screening and public education regarding thalassaemia should be emphasised through various mediums, such as campaigns and social media. These steps would build knowledge and awareness of this genetic disease in the society.

A rise in number of thalassaemia patients would be alarming. We hope to achieve a sustained reduction in new thalassaemia births with the implementation of the Form 4 thalassaemia screening programme. A carrier registry is very important to ensure that carriers can be identified early to prevent affected pregnancies. Pre-implantation genetic testing should also be readily available to allow carriers to have normal, disease-free children.

16 Terengganu

16.1 Introduction

The state of Terengganu has 6 districts, namely Besut, Setiu, Dungun, Hulu Terengganu, Kuala Terengganu and Kemaman. Terengganu has 6 hospitals from which thalassaemia patients seek treatment. Hospital Sultanah Nur Zahirah (HSNZ), Kuala Terengganu, being the state hospital and the only tertiary hospital in Terengganu, receives the largest number of thalassaemia patients.

Currently, apart from HSNZ, Hospital Kemaman and Hospital Hulu Terengganu also provides blood transfusion service as an out-patient treatment via day care centres. Although the other district hospitals, i.e. Hospital Dungun, Hospital Besut and Hospital Setiu, do not have day care centres, blood transfusion service for thalassaemia patients are delivered via day care concept. The patients no longer require overnight stay for transfusions, as what was previously practiced. This was made possible by collaboration with the state's blood bank.

16.2 Patient Demographics

Data analysed were taken from patients who are either living, lost to follow-up or cured by transplant. The total number of thalassaemia patients in Terengganu in 2019 is 345 patients, compared to 346 patients in 2018. There are 242 TDT patients and 103 NTDT patients. Table 16.1 summarises the number of patients in each centre in Terengganu, and Table 16.2 summarises the number of patients based on vital status.

Table 16.1: Distribution of Patients in Terengganu by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Sultanah Nur Zahirah, Kuala Terengganu	220	63.77
Hospital Dungun	23	6.67
Hospital Kemaman	34	9.86
Hospital Besut	23	6.67
Hospital Hulu Terengganu	34	9.86
Hospital Setiu	11	3.19
Total	345	100.00

Table 16.2: Distribution of Patients in Terengganu by Vital Status

Vital Status	Number of Patients
Alive and on active treatment	304
Cured by Stem Cell Therapy	1
Total	**305**
Lost to Follow-up	40
Total	**345**
Deaths in 2019	2
Cumulative Reported Deaths	28

There were 2 deaths of thalassaemia patients recorded in 2019 (up to November 2019). One patient who underwent haematopoietic stem cell transplantation (HSCT) in October 2017 has been cured, while another patient who underwent the procedure in October 2019 is still under observation. There were 4 new cases diagnosed in 2019: 1 patient from Hospital Kemaman and the remaining 3 patients from HSNZ, Kuala Terengganu.

Table 16.3: Cumulative Causes of Death in Terengganu

Causes of Death	Number of Patients	Percentage (%)
Infections	15	53.57
Cardiac Complications	8	28.57
Tumour	1	3.57
Thalassaemia	2	7.14
Liver Disease	1	3.57
Motor Vehicle Accident	1	3.57
Total	28	100.00

16.2.1 Age Group

Most thalassaemia patients in Terengganu are between the ages of 6-20 years old (185 out of 345 patients, 53.62%).

A majority of these patients are managed by the Paediatric Haemato-Oncologist based at HSNZ. Adult patients in HSNZ, on the other hand, are gradually transferred to adult haematologist.

Figure 16.1: Distribution of Patients in Terengganu by Age Group

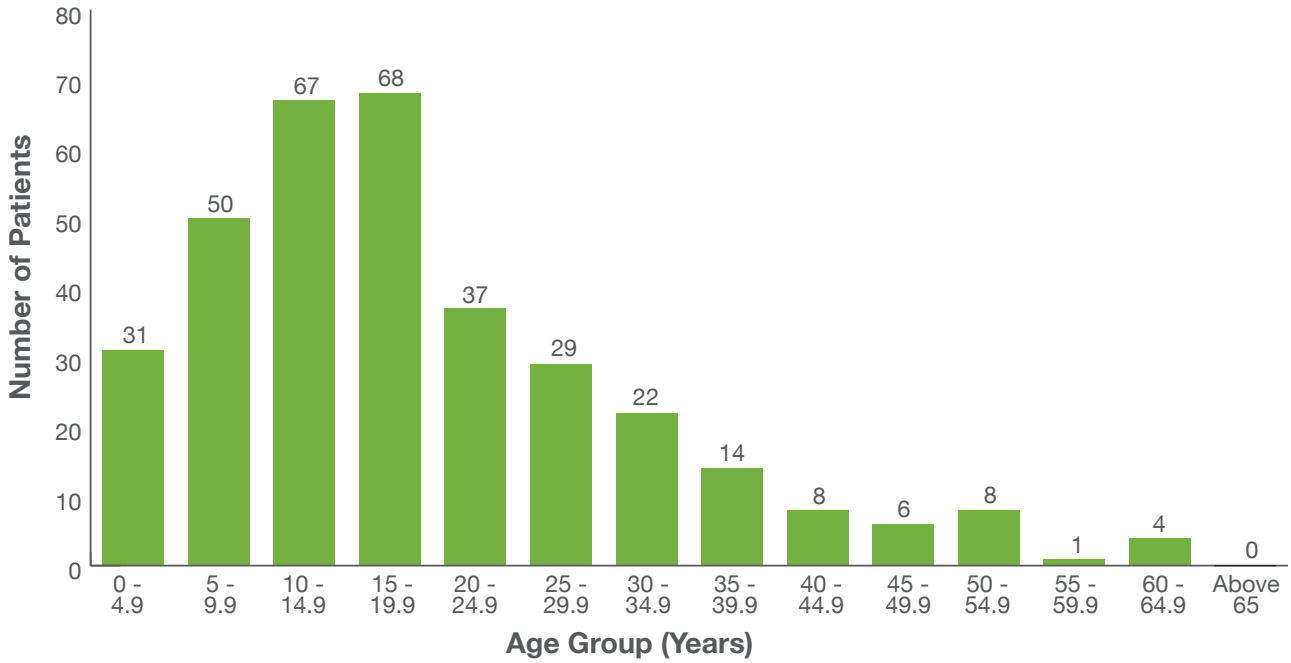


Table 16.4: Distribution of Patients in Terengganu According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	148	β-Thalassaemia Major	24	16.22
		β-Thalassaemia Intermedia	8	5.41
		HbE/β-Thalassaemia	79	53.38
		HbH Disease	34	22.97
		Others	3	2.03
15 - 29.9	134	β-Thalassaemia Major	29	21.64
		β-Thalassaemia Intermedia	8	5.97
		HbE/β-Thalassaemia	74	55.22
		HbH Disease	21	15.67
		Others	2	1.49
30 - 44.9	44	β-Thalassaemia Major	6	13.64
		β-Thalassaemia Intermedia	8	18.18
		HbE/β-Thalassaemia	27	61.36
		HbH Disease	2	4.55
		Others	1	2.27
45 - 59.9	15	β-Thalassaemia Major	2	13.33
		β-Thalassaemia Intermedia	5	33.33
		HbE/β-Thalassaemia	3	20.00
		HbH Disease	5	33.33
		Others	0	0.00
60 and above	4	β-Thalassaemia Major	1	25.00
		β-Thalassaemia Intermedia	1	25.00
		HbE/β-Thalassaemia	1	25.00
		HbH Disease	1	25.00
		Others	0	0.00
Total			345	

16.2.2 Gender

There are 164 female patients (47.54%) and 181 male patients (52.46%) in Terengganu. The distribution of male and female patients in each district hospital is shown in Table 16.5.

Table 16.5: Distribution of Patients in Terengganu According to Gender by Centre

Centre	Male		Female	
	No.	%	No.	%
Hospital Sultanah Nur Zahirah	116	33.62	104	30.14
Hospital Dungun	12	3.48	11	3.19
Hospital Kemaman	24	6.96	10	2.90
Hospital Besut	11	3.19	12	3.48
Hospital Hulu Terengganu	16	4.64	18	5.22
Hospital Setiu	2	0.58	9	2.61
Total	181	52.46	164	47.54

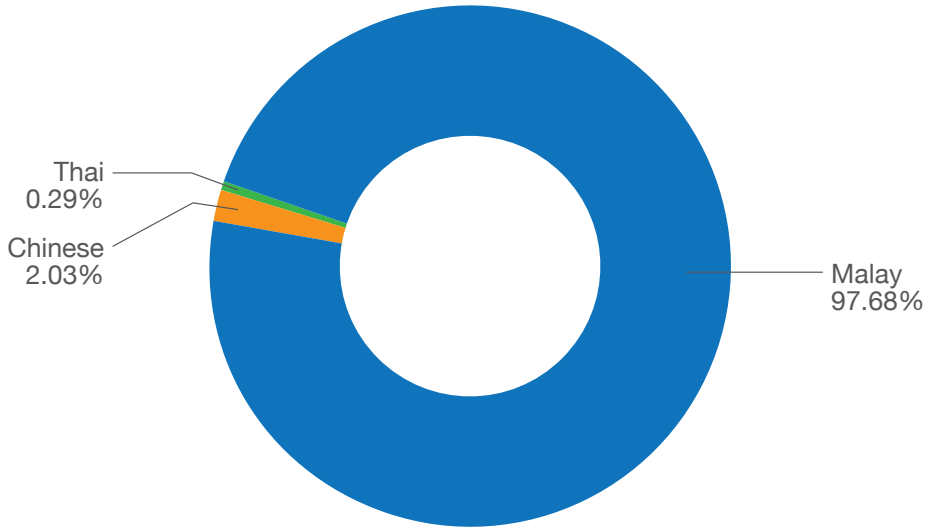
16.2.3 Ethnic Group

Table 16.6 shows the ethnic distribution of thalassaemia patients in Terengganu. Malay patients form a great majority of thalassaemia patients in the state (337 patients, 97.68%).

Table 16.6: Distribution of Patients in Terengganu According to Ethnic Group by Centre

Centre	Malay		Chinese		Thai	
	No.	%	No.	%	No.	%
Hospital Sultanah Nur Zahirah	214	62.03	6	1.74	0	0.00
Hospital Dungun	22	6.38	0	0.00	1	0.29
Hospital Kemaman	33	9.57	1	0.29	0	0.00
Hospital Besut	23	6.67	0	0.00	0	0.00
Hospital Hulu Terengganu	34	9.86	0	0.00	0	0.00
Hospital Setiu	11	3.19	0	0.00	0	0.00
Total	337	97.68	7	2.03	1	0.29

Figure 16.2: Distribution of Patients in Terengganu by Ethnic Group



16.3 Diagnosis

As shown in Figure 16.3, HbE/β-thalassaemia is the main diagnosis of the patients in Terengganu (184 patients, 53.33%), followed by HbH disease (63 patients, 18.26%) and β-thalassaemia major (62 patients, 17.97%). HbE/β-thalassaemia diagnosis is prominent in all 5 centres located in Terengganu, except in Hospital Hulu Terengganu, which reported β-thalassaemia major as the most frequent diagnosis (Table 16.7).

Figure 16.3: Distribution of Patients in Terengganu by Diagnosis

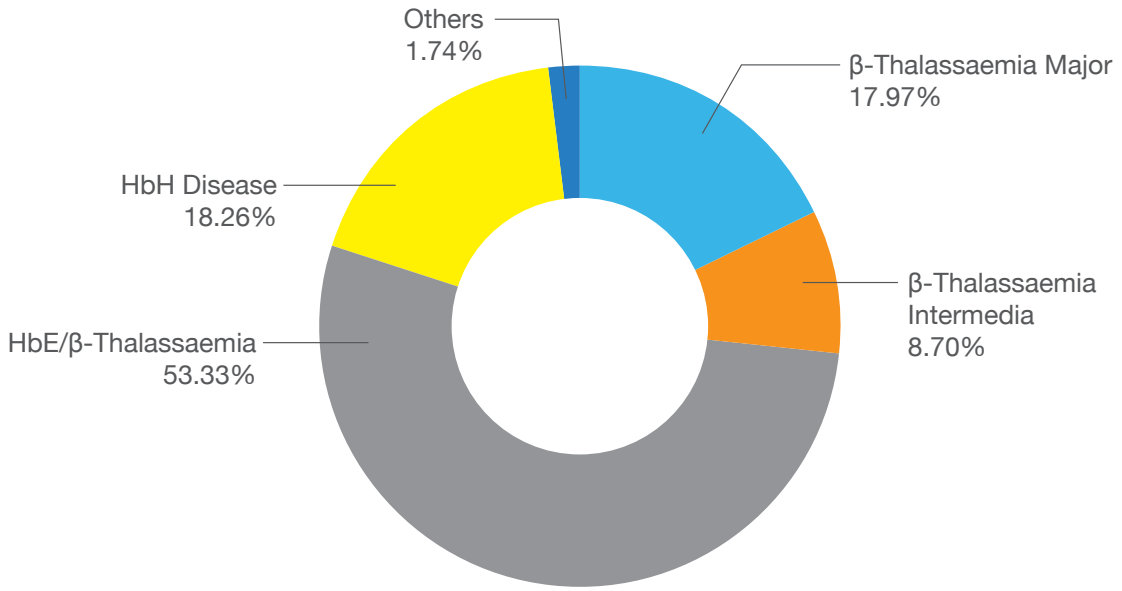


Table 16.7: Distribution of Patients in Terengganu According to Diagnosis by Centre

Centre	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/ β-Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Nur Zahirah	25	7.25	20	5.80	123	35.65	48	13.91	4	1.16
Hospital Dungun	7	2.03	1	0.29	14	4.06	1	0.29	0	0.00
Hospital Kemaman	5	1.45	3	0.87	21	6.09	4	1.16	1	0.29
Hospital Besut	8	2.32	2	0.58	9	2.61	4	1.16	0	0.00
Hospital Hulu Terengganu	16	4.64	4	1.16	9	2.61	4	1.16	1	0.29
Hospital Setiu	1	0.29	0	0.00	8	2.32	2	0.58	0	0.00
Total	**62**	**17.97**	**30**	**8.70**	**184**	**53.33**	**63**	**18.26**	**6**	**1.74**

Table 16.8: Distribution of Patients in Terengganu According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	62	Malay	60	17.39
		Chinese	1	0.29
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	1	0.29
β-Thalassaemia Intermedia	30	Malay	28	8.12
		Chinese	2	0.58
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
HbE/ β-Thalassaemia	184	Malay	183	53.04
		Chinese	1	0.29
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
HbH Disease	63	Malay	60	17.39
		Chinese	3	0.87
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Others	6	Malay	6	1.74
		Chinese	0	0.00
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Total			345	

16.4 Treatment

16.4.1 Iron Chelation Therapy

A total of 239 thalassaemia patients (69.27%) in Terengganu are receiving chelating agents. As shown in Table 16.9, DFP is the most commonly prescribed agent (111 patients out of 239, 46.44%), followed by DFX (74 patients, 30.96%) and DFO (24 patients, 10.04%). All centres in Terengganu mostly utilise DFP, except for Hospital Kemaman (Table 16.10).

Table 16.9: Distribution of Patients in Terengganu by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	30	12.55
DFP only	111	46.44
DFX only	74	30.96
DFO + DFP	24	10.04
DFP + DFX	0	0.00
DFO + DFX	0	0.00
DFO + DFP + DFX	0	0.00
Total	239	100.00

Table 16.10: Distribution of Patients in Terengganu According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Sultanah Nur Zahirah	147	DFO only	23	9.62
		DFP only	58	24.27
		DFX only	44	18.41
		DFO + DFP	22	9.21
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Dungun	21	DFO only	1	0.42
		DFP only	13	5.44
		DFX only	6	2.51
		DFO + DFP	1	0.42
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kemaman	23	DFO only	0	0.00
		DFP only	8	3.35
		DFX only	15	6.28
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Besut	13	DFO only	3	1.26
		DFP only	7	2.93
		DFX only	3	1.26
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Hulu Terengganu	27	DFO only	2	0.84
		DFP only	19	7.95
		DFX only	5	2.09
		DFO + DFP	1	0.42
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Setiu	8	DFO only	1	0.42
		DFP only	6	2.51
		DFX only	1	0.42
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			239	100.00

Table 16.11: Distribution of Patients in Terengganu According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0–14.9	100	DFO only	11	11.00
		DFP only	26	26.00
		DFX only	62	62.00
		DFO + DFP	1	1.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15–29.9	96	DFO only	15	15.63
		DFP only	58	60.42
		DFX only	8	8.33
		DFO + DFP	15	15.63
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30–44.9	33	DFO only	3	9.09
		DFP only	20	60.61
		DFX only	3	9.09
		DFO + DFP	7	21.21
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45–59.9	8	DFO only	1	12.50
		DFP only	6	75.00
		DFX only	0	0.00
		DFO + DFP	1	12.50
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

60 and above	2	DFO only	0	0.00
		DFP only	1	50.00
		DFX only	1	50.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			239	

Chelating agents are mostly prescribed to patients below 30 years old. DFX is mainly prescribed to patients between 0–10 years old, whereas DFP is mostly prescribed to patients aged 11 years and above.

16.4.2 Serum Ferritin Level

Out of 242 TDT patients, 222 (91.74%) have had their serum ferritin levels recorded. A total of 144 patients (59.50% out of 242 TDT patients) have a serum ferritin level lower than 2500 ng/mL. The remaining 78 patients (32.23%) have a serum ferritin level higher than 2500 ng/mL, including 3 patients with serum ferritin levels exceeding 10,000 ng/mL.

Table 16.12: Distribution of Patients in Terengganu According to Most Recent Serum Ferritin Level by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Nur Zahirah	145	53	23.87	47	21.17	31	13.96	14	6.31	0	0.00
Hospital Dungun	11	2	0.90	7	3.15	2	0.90	0	0.00	0	0.00
Hospital Kemaman	24	3	1.35	12	5.41	6	2.70	3	1.35	0	0.00
Hospital Besut	8	3	1.35	0	0.00	1	0.45	4	1.80	0	0.00
Hospital Hulu Terengganu	29	6	2.70	10	4.50	6	2.70	4	1.80	3	1.35
Hospital Setiu	5	0	0.00	1	0.45	3	1.35	1	0.45	0	0.00
Total	222	67	30.18	77	34.68	49	22.07	26	11.71	3	1.35

16.5 Conclusion

A majority of thalassaemia patients in Terengganu receive treatment at a tertiary centre, i.e. HSNZ (220 out of 345 patients, 63.77%). Patients aged 20 years and below form the largest group with 216 patients (62.61%). Nearly all thalassaemia patients in Terengganu are of Malay ethnicity (337 patients, 97.68%). Male to female patient ratio is 1:1. According to the current registry data, 4 new cases were reported in 2019.

HbE/β-thalassaemia is the main diagnosis of the patients in Terengganu (184 patients, 53.33%), followed by HbH disease (63 patients, 18.26%) and β-thalassaemia major (62 patients, 17.97%). There were 2 deaths of thalassaemia patients in 2019, bringing to a cumulative total of 28 deaths recorded in the state. One patient underwent HSCT treatment at IPHKL in October 2017, and is now considered cured. Another patient who underwent HSCT is still under observation.

DFP is the most commonly prescribed chelating agent, taken by 111 patients (46.44%), followed by DFX with 74 patients (30.96%). DFO + DFP is the most common combination of chelating agents prescribed in Terengganu. Patients who are not on chelating agents are mostly of the NTDT type. In addition, 144 patients out of 242 TDT patients (59.50%) in Terengganu have serum ferritin levels lower than 2500 ng/mL.

17 Wilayah Persekutuan Kuala Lumpur

17.1 Introduction

The federal territories comprise 3 territories, which are Kuala Lumpur, Putrajaya and Labuan. Both Kuala Lumpur and Putrajaya are enclaves in the state of Selangor, whereas Labuan is an island off the coast of the state of Sabah.

Kuala Lumpur is the capital of Malaysia. The city proper, making up an area of 243 km^2 (94 sq mi), has a population of 1.8 million as of 2019. The ethnic distribution in Kuala Lumpur are as follows: Malay (40.19%), Chinese (36.55%), Indian (8.46%), and others (14.80%).

There are 3 major public hospitals in Kuala Lumpur: Institute of Paediatrics, Kuala Lumpur Hospital (IPHKL), Universiti Kebangsaan Malaysia Medical Centre (PPUKM), and Universiti Malaya Medical Centre (PPUM). Paediatric services were gradually moved from the Institute of Paediatrics, Kuala Lumpur to Hospital Wanita dan Kanak-Kanak Kuala Lumpur on 17th of April 2019. The move was completed on 13th of September 2019. These 3 hospitals are the referral centres for paediatric diseases, including haematological disorders such as thalassaemia. The centres which perform allogeneic stem cell transplant for thalassaemia patients in Kuala Lumpur are IPHKL, PPUM and Subang Jaya Medical Centre (SJMC).

Prior to the national thalassaemia screening and prevention programme, an estimated 40% of patients are already on some form of iron chelation therapy, although adequacy and compliance may not be ideal. With the availability of iron chelation therapy, initiatives by the government are now focused on achieving ideal iron levels in the body and preventing long-term morbidities.

17.2 Patient Demographics

Patients were categorised as living, lost to follow-up, cured by transplant or deceased. The cumulative number of thalassaemia patient deaths in Kuala Lumpur remained at 90 since 1997 until 2019. There are 506 thalassaemia patients in Kuala Lumpur, including 99 patients lost to follow-up and 33 patients cured by transplant.

Table 17.1: Distribution of Patients in Kuala Lumpur by Centre

Centre	Number of Patients (n)	Percentage (%)
IPHKL	214	42.29
PPUKM	180	35.57
PPUM	112	22.13
Total	506	100.00

Table 17.2: Distribution of Patients in Kuala Lumpur by Vital Status

Status	Number of Patients
Alive and On Active Treatment	374
Cured by Stem Cell Therapy	33
Total	**407**
Lost to Follow-up	99
Total	**506**
Deaths in 2019	0
Cumulative Reported Deaths	90

Table 17.3: Cumulative Causes of Death in Kuala Lumpur Since 1997

Causes of Death	Number of Patients	Percentage (%)
Infections	25	37.31
Cardiac Complications	35	52.24
Endocrine Complications	1	1.49
Others	3	4.48
Tumours	1	1.49
Central Nervous System Event	1	1.49
Motor Vehicle Accident	1	1.49
Total	67	100.00

Twenty-three deceased patients had incomplete data; these patients' causes of death are not included in the table.

17.2.1. Age Group

The youngest patient in Kuala Lumpur is 11 months old and the oldest is 66 years old. As shown in Figure 17.1, patients aged 5-19.9 years old form the largest group of patients in Kuala Lumpur. Of these, a sizeable number was transferred to the adult treatment centre once they reached 18 years of age (based on year of birth). Table 17.4 shows that most patients aged below 45 years old are diagnosed with HbE/β-thalassaemia. Only 1 patient in the 45-59.9 years age group is diagnosed with β-thalassaemia intermedia, and 1 patient aged above 60 years old is diagnosed with β-thalassaemia major.

Figure 17.1: Distribution of Patients in Kuala Lumpur by Age Group

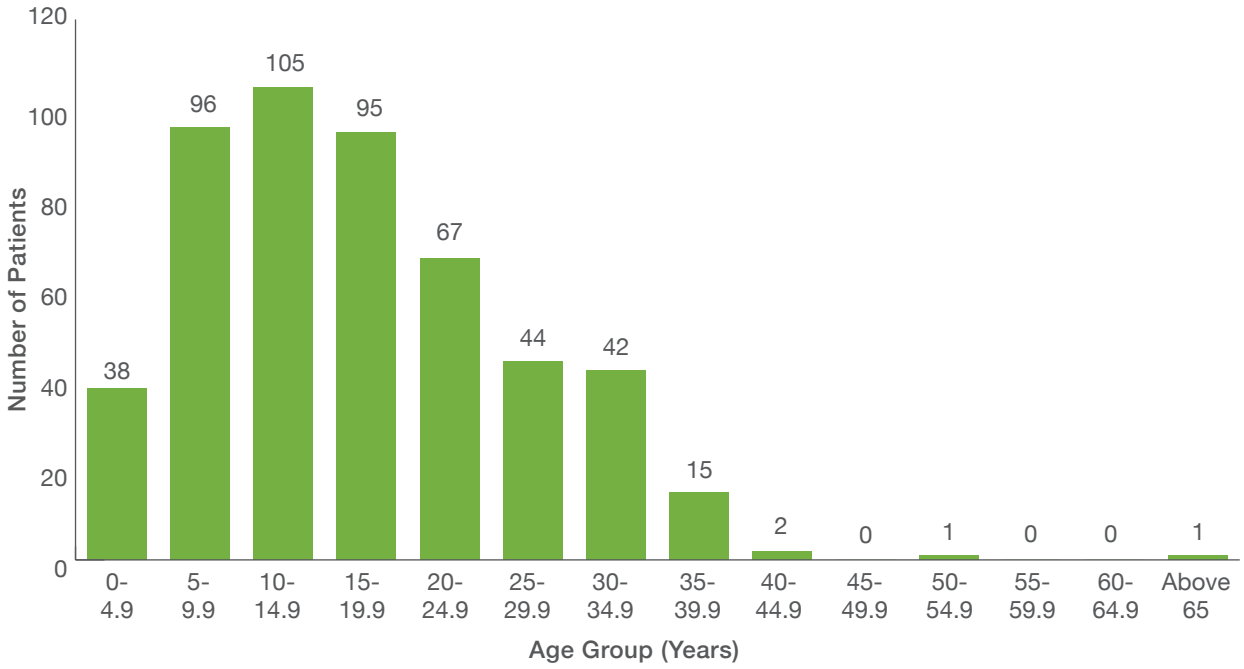


Table 17.4: Distribution of Patients in Kuala Lumpur According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	239	β-Thalassaemia Major	60	25.10
		β-Thalassaemia Intermedia	7	2.93
		HbE/β-Thalassaemia	105	43.93
		HbH Disease	45	18.83
		Others	22	9.21
15 - 29.9	206	β-Thalassaemia Major	56	27.18
		β-Thalassaemia Intermedia	5	2.43
		HbE/β-Thalassaemia	95	46.12
		HbH Disease	45	21.84
		Others	5	2.43
30 - 44.9	59	β-Thalassaemia Major	19	32.20
		β-Thalassaemia Intermedia	3	5.08
		HbE/β-Thalassaemia	26	44.07
		HbH Disease	10	16.95
		Others	1	1.69
45 - 59.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	100.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
60 and above	1	β-Thalassaemia Major	1	100.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			506	

17.2.2. Gender

The gender distribution of thalassaemia patients in Kuala Lumpur is almost equal. There are 268 male patients (52.96%) and 238 female patients (47.04%) in Kuala Lumpur (Table 17.5).

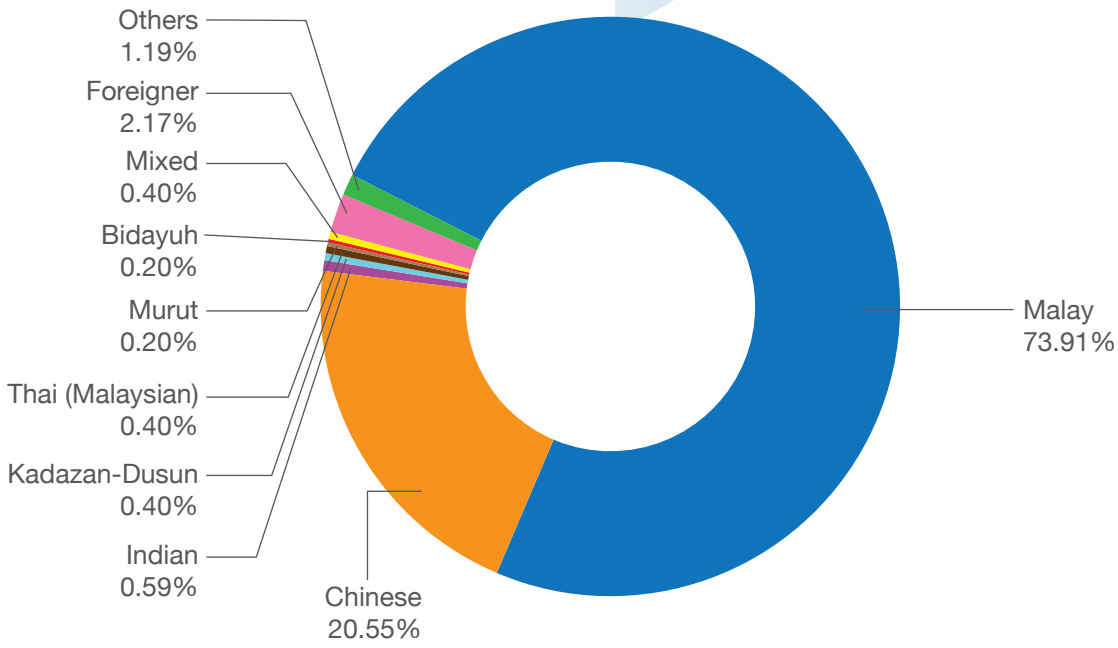
Table 17.5: Distribution of Patients in Kuala Lumpur According to Gender by Centre

Centre	Male		Female	
	No.	%	No.	%
IPHKL	113	22.33	101	19.96
PPUKM	98	19.37	82	16.21
PPUM	57	11.26	55	10.87
Total	268	52.96	238	47.04

17.2.3. Ethnic Group

Malay patients form the majority (374 patients, 73.91%) of patients in Kuala Lumpur, followed by the Chinese with 104 patients (20.55%) and Indian with 3 patients (0.59%). Eleven non-Malaysian patients are also seeking treatment in Kuala Lumpur. For the non-citizens, the treatment fees are as per the rate for foreigners, with self-purchase of iron chelators (Figure 17.2).

Figure 17.2: Distribution of Patients in Kuala Lumpur by Ethnic Group



17.3 Diagnosis

According to Figure 17.3, the diagnoses of HbE/β-thalassaemia and β-thalassaemia major contribute to 44.66% (226 patients) and 26.88% (136 patients) of the patients in Kuala Lumpur, respectively. HbH disease contributes to 19.76% of patients (100 patients). β-thalassaemia intermedia contributes to 3.16%, and the remaining 5.53% (28 patients) have other diagnoses. HbE/β-thalassaemia is the most prevalent type of diagnosis among the Malay, whereas β-thalassaemia major is prevalent among Chinese (Table 17.6).

Figure 17.3: Distribution of Patients in Kuala Lumpur by Diagnosis

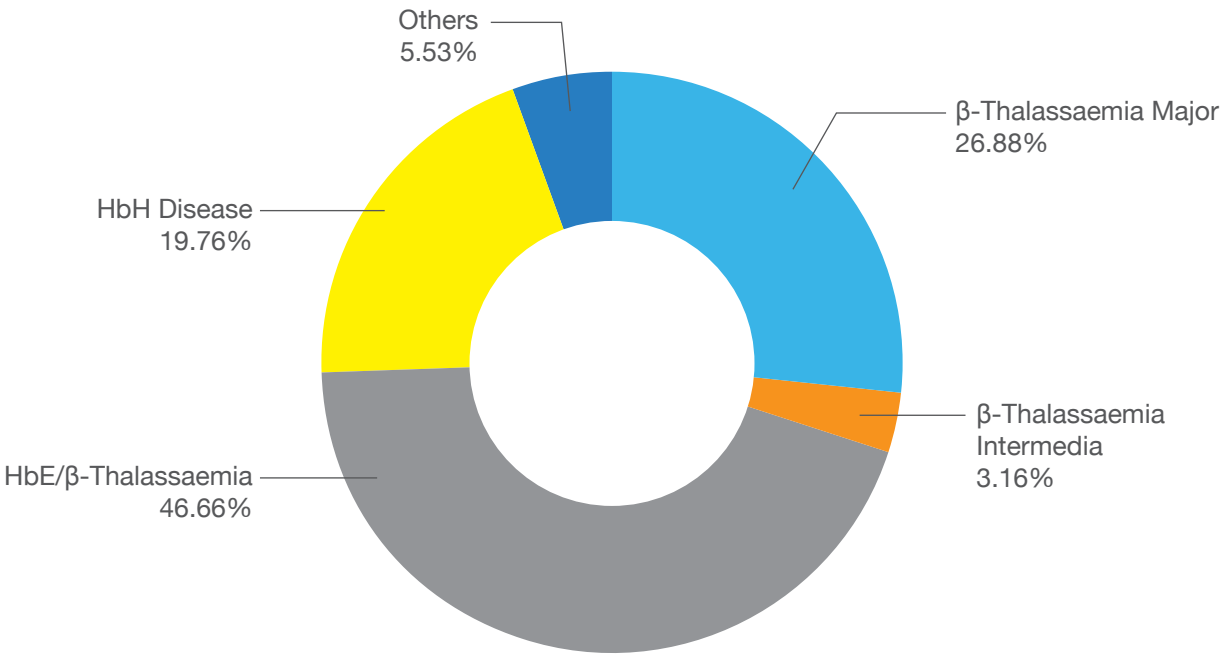


Table 17.6: Distribution of Patients in Kuala Lumpur According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	136	Malay	67	13.24
		Chinese	56	11.07
		Indian	1	0.20
		Kadazan-Dusun	1	0.20
		Others	11	2.17
β-Thalassaemia Intermedia	16	Malay	10	1.98
		Chinese	4	0.79
		Indian	1	0.20
		Kadazan-Dusun	0	0.00
		Others	1	0.20
HbE/ β-Thalassaemia	226	Malay	199	39.33
		Chinese	15	2.96
		Indian	0	0.00
		Kadazan-Dusun	1	0.20
		Others	11	2.17
HbH Disease	100	Malay	73	14.43
		Chinese	27	5.34
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Others	28	Malay	25	4.94
		Chinese	2	0.40
		Indian	1	0.20
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Total			506	100.00

*

17.4 Treatment

17.4.1 Iron Chelation Therapy

Currently, there are 293 TDT and NTDT patients (57.91%) receiving iron chelation therapy in Kuala Lumpur. The remaining 213 (42.09%) patients do not receive chelation therapy. Of the patients receiving iron chelation therapy, 232 patients (79.18%) are on monotherapy and 61 patients (20.82%) are on a combination of iron chelators (Table 17.7). The main combination of chelators used is DFO + DFP, prescribed to 48 patients (16.38%). The type of iron chelator received by patients vary according to age groups. As shown in Table 17.9, a majority of patients below 15 years old receive DFX monotherapy. Meanwhile, the 15-29.9 and 30-44.5 years age groups have the highest number of patients on DFO and DFP monotherapy, respectively. The oldest patient in Kuala Lumpur is aged above 60 years old, and receives a combination of DFO and DFP therapy.

Table 17.7: Distribution of Patients in Kuala Lumpur by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	58	19.80
DFP only	38	12.97
DFX only	136	46.42
DFO + DFP	48	16.38
DFP + DFX	3	1.02
DFO + DFX	10	3.41
DFO + DFP + DFX	0	0.00
Total	293	100.00

Table 17.8: Distribution of Patients in Kuala Lumpur According to Type of Iron Chelator Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
IPHKL	106	DFO only	11	3.75
		DFP only	1	0.34
		DFX only	83	28.33
		DFO + DFP	2	0.68
		DFP + DFX	0	0.00
		DFO + DFX	9	3.07
		DFO + DFP + DFX	0	0.00
PPUKM	89	DFO only	10	3.41
		DFP only	19	6.48
		DFX only	35	11.95
		DFO + DFP	23	7.85
		DFP + DFX	2	0.68
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
PPUM	98	DFO only	37	12.63
		DFP only	18	6.14
		DFX only	18	6.14
		DFO + DFP	23	7.85
		DFP + DFX	1	0.34
		DFO + DFX	1	0.34
		DFO + DFP + DFX	0	0.00
Total			293	100.00

Table 17.9: Distribution of Patients in Kuala Lumpur According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0–14.9	145	DFO only	17	11.72
		DFP only	6	4.14
		DFX only	111	76.55
		DFO + DFP	4	2.76
		DFP + DFX	0	0.00
		DFO + DFX	7	4.83
		DFO + DFP + DFX	0	0.00
15–29.9	114	DFO only	33	28.95
		DFP only	21	18.42
		DFX only	24	21.05
		DFO + DFP	32	28.07
		DFP + DFX	1	0.88
		DFO + DFX	3	2.63
		DFO + DFP + DFX	0	0.00
30–44.9	32	DFO only	8	25.00
		DFP only	10	31.25
		DFX only	1	3.13
		DFO + DFP	11	34.38
		DFP + DFX	2	6.25
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45–59.9	1	DFO only	0	0.00
		DFP only	1	100.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	100.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			293	

17.4.2 Serum Ferritin Level

There are 254 TDT patients recorded in Kuala Lumpur in 2018, and 247 TDT patients recorded in 2019 (Table 17.10). Forty-eight patients (18.90%) have a serum ferritin level lower than 1000 ng/mL in 2018, which is reduced to 45 patients (18.22%) in 2019. A total of 101 patients (39.76%) and 105 patients (42.51%) have a serum ferritin level between 1000-2500 ng/mL in 2018 and 2019, respectively. On the other hand, the number of patients with serum ferritin levels above 2500 ng/mL was 105 (41.34%) in 2018 and 97 (39.27%) in 2019.

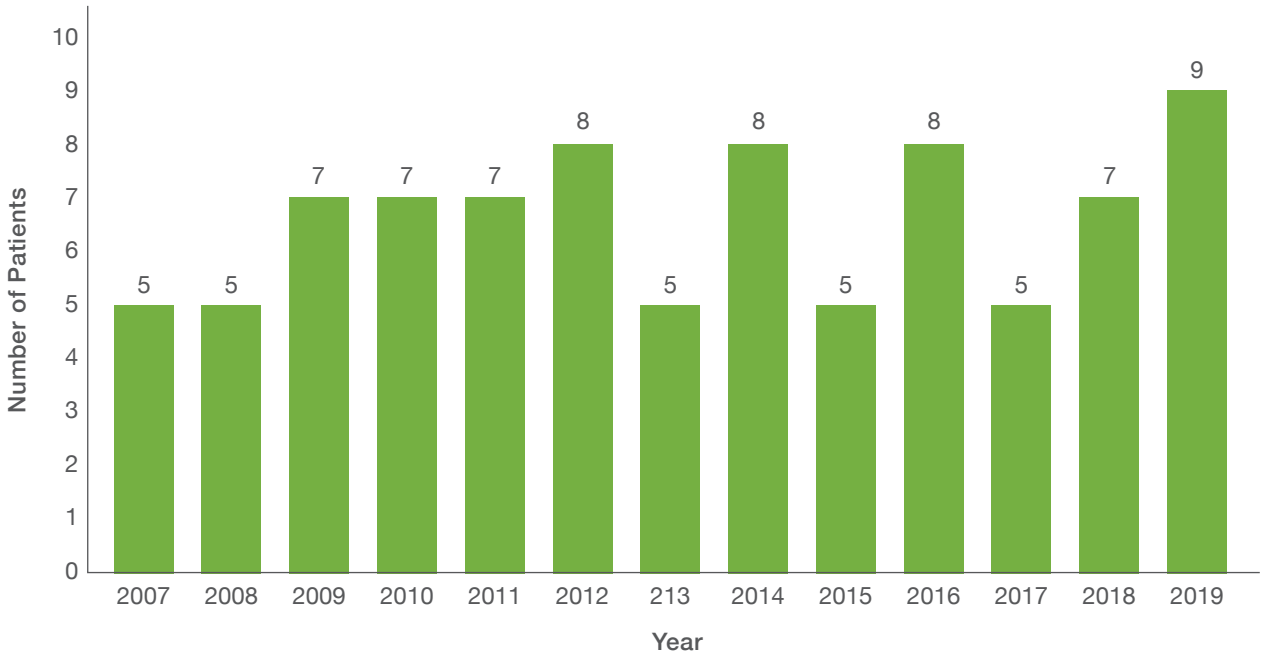
Table 17.10: Distribution of Patients in Kuala Lumpur According to Serum Ferritin Level in 2019 by Centre

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
IPHKL	99	20	8.10	49	19.84	21	8.50	8	3.24	1	0.40
PPUKM	64	15	6.07	18	7.29	21	8.50	10	4.05	0	0.00
PPUM	84	10	4.05	38	15.38	24	9.72	11	4.45	1	0.40
Total	247	45	18.22	105	42.51	66	26.72	29	11.74	2	0.81

17.4.3 Haemopoietic Stem Cell Transplant in Thalassaemia

A total of 86 patients underwent stem cell transplant in IPHKL since 2007. The patients were referred from hospitals throughout Malaysia.

Figure 17.4: Number of Thalassaemia Patients Who Underwent Stem Cell Transplant at IPHKL by Year



17.5 Conclusion

This report focuses on the results of patient management and outcomes ever since the MTR has been operational, i.e. from 2007 to 2019. It describes the outcomes based on demographic statistics, the types of thalassaemia and effectiveness of treatment.

There are 268 (52.96%) male and 238 (47.04%) female patients in Kuala Lumpur. The youngest patient is 11 months old and the oldest is 66 years old. Patients aged between 10-15 years form the largest age group of patients in Kuala Lumpur. Generally, as paediatric patients turned 18 years old, they will be transferred to one of the adult treatment centres (usually Hospital Ampang) or adult haematology clinics in PPUKM and PPUM. Malay patients form the largest group with 374 patients (73.91%), followed by Chinese with 104 patients (20.55%) and Indian with only 3 patients (0.59%). A total of 11 foreigners are also seeking treatment for thalassaemia in Kuala Lumpur. The treatment fees and cost of iron chelators are self-paid by foreigners; thus, the cost of treatment is higher compared to the citizens.

Most children with moderate to severe thalassaemia show signs and symptoms within the first two years of life. A diagnosis of thalassaemia is confirmed by Hb analysis that uses high-performance liquid chromatography (HPLC) test and capillary electrophoresis (CE). HbE/β-thalassaemia and β-thalassaemia major afflicted 226 (44.66%) and 136 (26.88%) patients in Kuala Lumpur, respectively. HbH disease contributes 100 patients (19.76%), whereas 28 (5.53%) patients have other diagnoses.

Iron chelation therapy is essential for patients with thalassaemia to reduce iron overload. In total, 232 patients (79.18%) in Kuala Lumpur are on monotherapy and 61 patients (20.82%) are on combination therapy. The most common combination of chelators is DFO + DFP, prescribed to 48 patients (16.38%). A significant number of patients are on DFX monotherapy (136 patients, 46.42%).

A total of 149 out of 254 patients (58.66%) had a serum ferritin level lower than 2500 ng/mL in 2018, which raised 150 out of 247 patients (60.73%) in 2019. It can be concluded that the number of patients who are compliant to iron chelation therapy was slightly increased in 2019.

18 Wilayah Persekutuan Labuan

18.1 Introduction

Labuan is a federal territory located in in East Malaysia. It is an island off the shoreline of the territory of Sabah. The Federal Territory of Labuan comprises Labuan Island (75 km^2) and 6 other smaller islands (Pulau Burung, Pulau Daat, Pulau Kuraman, Pulau Papan, Pulau Rusukan Kecil, and Pulau Rusukan Besar), which have a combined total area of 91 km^2.

Labuan has a population of 99,200 as of 2018 (Department of Statistics Malaysia). The population is comprised of Bumiputera (75,000, 75.60%), Chinese (9,500, 9.58%), Indian (800, 0.80%), other ethnicities (2,800, 2.82%) and non-Malaysian citizens (11,100, 11.19%).

Hospital Labuan is the only centre in Labuan. The development of Hospital Labuan was started on the 10th of October 1991. It is situated on an island close to the state of Sabah, in an area of 12,047 hectares and a building site area of around 31,969 m^2. Its features are not built like other hospitals because of the building concept nucleus as figure. Hospital Labuan is situated 6 miles from downtown Federal Territory of Labuan, right amidst the island.

Hospital Labuan commenced operations on the 10th of June 1995, and was inaugurated by former Prime Minister Tun Dr. Mahathir bin Mohamad on the 29th of August 1996. It has a capacity of 109 beds. Hospital Labuan has a specialist physician, surgeon general and anaesthetist experts for disciplines such as ear, nose, and throat (ENT), paediatrics, and psychiatry. All specialist conduct thalassaemia care in the clinics.

18.2 Patient Demographics

There are 27 living thalassaemia patients in Labuan, including 1 patient who underwent stem cell therapy (Tables 18.1 and 18.2). The cumulative number of deaths of thalassaemia patients in Labuan since 2007 is 4 patients. Two of these patients had incomplete data, hence their causes of death are unknown (Table 18.3 and 18.4).

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Table 18.1: Distribution of Patients in Labuan by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Labuan	27	100.00
Total	27	100.00

Table 18.2: Distribution of Patients in Labuan by Vital Status

Status	Number of Patients
Alive and On Active Treatment	26
Cured by Stem Cell Therapy	1
Total	**27**
Lost to Follow-up	0
Total	**27**
Deaths in 2019	0
Cumulative Reported Deaths	4

Table 18.3: Cumulative Causes of Death in Labuan Since 2007

Causes of Death	Number of Patients	Percentage (%)
Infection	1	50.00
Other	1	50.00
Total	2	100.00

There have been 4 deaths of thalassaemia patients in Labuan since 2007. Two of these patients had incomplete data, hence their causes of death are unknown.

18.2.1. Age Group

As shown Figure 18.1, 26 out of the 27 patients in Labuan are aged below 25 years old. Most of these patients are diagnosed with β-thalassaemia major (Table 18.4). The remaining 1 patient falls in the 45-49.9 years age group, and is afflicted with β-thalassaemia intermedia.

Figure 18.1: Distribution of Patients in Labuan by Age Group

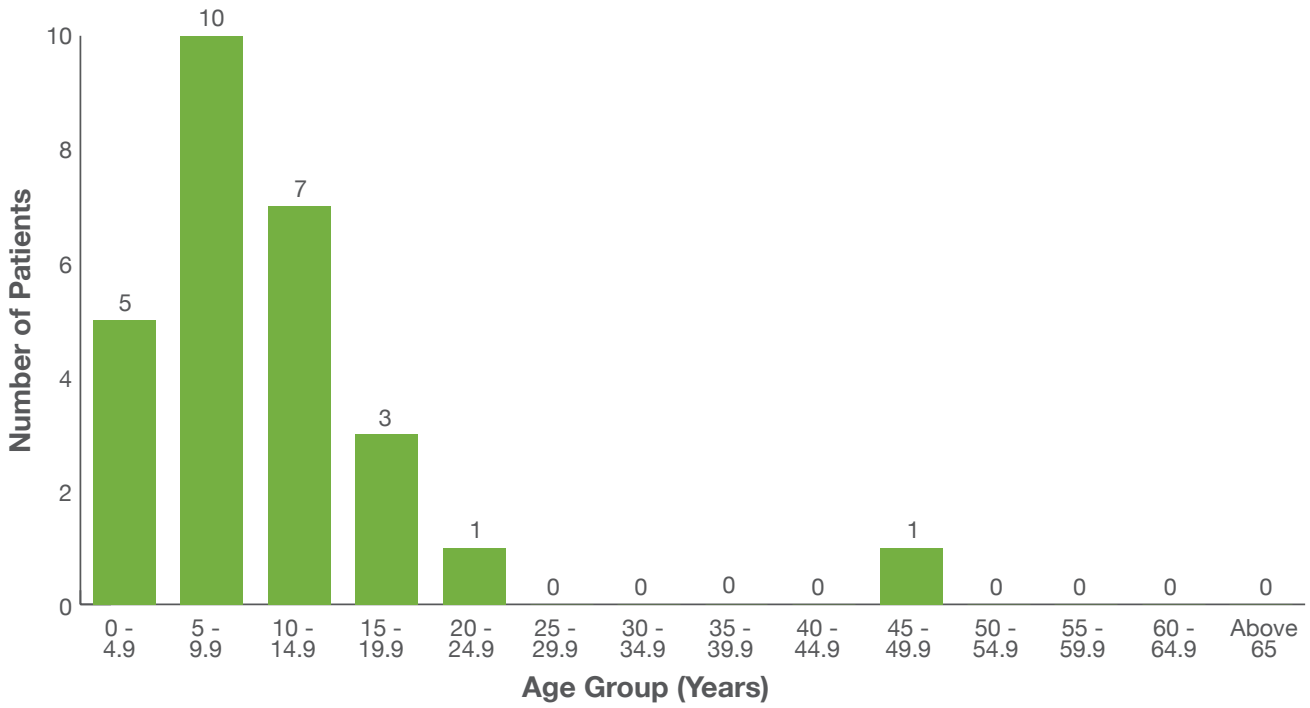


Table 18.4: Distribution of Patients in Labuan According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	22	β-Thalassaemia Major	12	54.55
		β-Thalassaemia Intermedia	7	31.82
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	4.55
		Others	2	9.09
15 - 29.9	4	β-Thalassaemia Major	3	75.00
		β-Thalassaemia Intermedia	1	25.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00

30 - 44.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
45 - 59.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	100.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			27	

18.2.2 Gender

Table 18.5 shows the distribution of thalassaemia patients in Labuan by gender. Female patients are higher in number with 17 patients (62.96%), compared to only 10 male patients (37.04%).

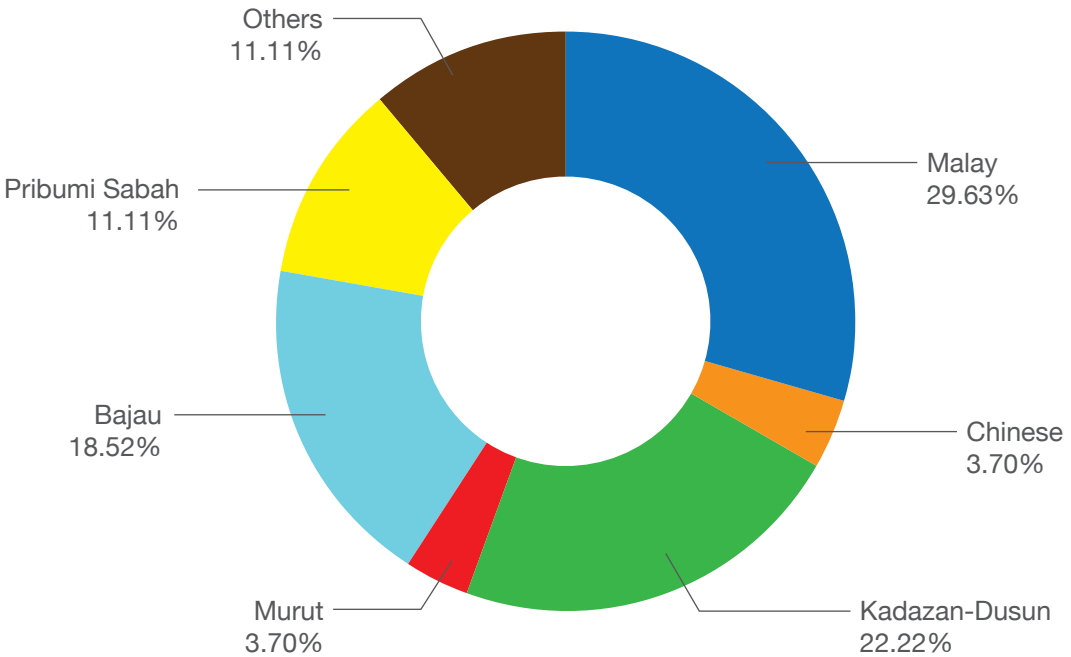
Table 18.5: Distribution of Patients in Labuan by Gender

Centre	Male		Female	
	No.	%	No.	%
Hospital Labuan	10	37.04	17	62.96
Total	10	37.04	17	62.96

18.2.3 Ethnic Group

The Malay forms the largest group of patients in Labuan with 8 patients (29.63%), followed by Kadazan-Dusun with 6 patients (22.22%) and Bajau with 5 patients (18.52%). Pribumi Sabah and Other ethnicities contribute 3 patients (11.11%) each. Finally, the Chinese and Murut contribute 1 patient (3.70%) each. The distribution of thalassaemia patients in Labuan according to ethnicity is shown in Figure 18.2.

Figure 18.2: Distribution of Patients in Labuan by Ethnic Group



18.3 Diagnosis

Most patients (15 patients, 55.56%) in Labuan have β-thalassaemia major, followed by β-thalassaemia intermedia (9 patients, 33.33%). One patient (3.70%) has HbH disease, and the remaining 2 patients (7.41%) have other diagnoses, as shown in Figure 18.3. The Kadazan-Dusun contributes the highest number of β-thalassaemia major patients, whereas the Malay has the highest number of β-thalassaemia intermedia patients (Table 18.6).

Figure 18.3: Distribution of Patients in Labuan by Diagnosis

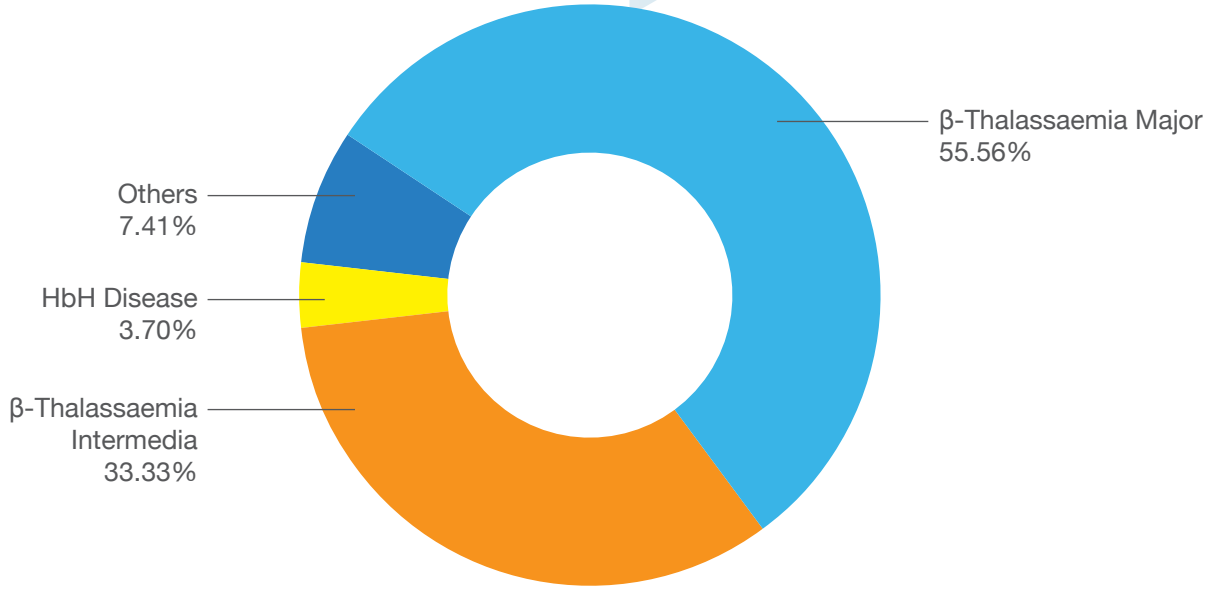


Table 18.6: Distribution of Patients in Labuan According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnic Group	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	15	Malay	4	14.81
		Chinese	0	0.00
		Kadazan-Dusun	5	18.52
		Murut	1	3.70
		Bajau	3	11.11
		Pribumi Sabah	0	0.00
		Others	2	7.41
β-Thalassaemia Intermedia	9	Malay	4	14.81
		Chinese	1	3.70
		Kadazan-Dusun	1	3.70
		Murut	0	0.00
		Bajau	1	3.70
		Pribumi Sabah	2	7.41
		Others	0	0.00
HbE/β-Thalassaemia	0	Malay	0	0.00
		Chinese	0	0.00
		Kadazan-Dusun	0	0.00
		Murut	0	0.00
		Bajau	0	0.00
		Pribumi Sabah	0	0.00
		Others	0	0.00

HbH Disease	1	Malay	0	0.00
		Chinese	0	0.00
		Kadazan-Dusun	0	0.00
		Murut	0	0.00
		Bajau	1	3.70
		Pribumi Sabah	0	0.00
		Others	0	0.00
Others	2	Malay	0	0.00
		Chinese	0	0.00
		Kadazan-Dusun	0	0.00
		Murut	0	0.00
		Bajau	0	0.00
		Pribumi Sabah	1	3.70
		Others	1	3.70
Total			27	100.00

18.4 Treatment

18.4.1 Iron Chelation Therapy

Only 18 out of 27 patients in Labuan receive iron chelation therapy. Nine patients (50%) are on DFX, 3 patients (16.67%) each are on DFO and DFO + DFP combination, 2 patients (11.11%) are on DFP + DFX combination and 1 patient (5.56%) is on DFO + DFP + DFX combination (Table 18.7). All patients on DFX monotherapy are aged below 15 years old. Two out of 4 patients in 15-29.9 years age group are on DFO monotherapy, whereas the remaining 2 patients are each receiving a combination therapy of DFO + DFP and DFP + DFX, respectively. The oldest patient in the age group of 45-59.9 years receive a combination therapy of DFO + DFP (Table 18.8).

Table 18.7: Distribution of Patients in Labuan by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	3	16.67
DFP only	0	0.00
DFX only	9	50.00
DFO + DFP	3	16.67
DFP + DFX	2	11.11
DFO + DFX	0	0.00
DFO + DFP + DFX	1	5.56
Total	18	100.00

Table 18.8: Distribution of Patients in Labuan According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	13	DFO only	1	7.69
		DFP only	0	0.00
		DFX only	9	69.23
		DFO + DFP	1	7.69
		DFP + DFX	1	7.69
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	7.69
15 - 29.9	9	DFO only	2	50.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	25.00
		DFP + DFX	1	25.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 44.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 59.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	100.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			18	

18.4.2 Serum Ferritin Level

Only 18 patients in Labuan have had their serum ferritin levels measured in 2019 (Table 18.9). The lowest serum ferritin level recorded is 107.50 ng/mL, and the highest is 19,483.00 ng/mL. Five patients (27.78%) have serum ferritin levels lower than 1000 ng/mL, 2 patients (11.11%) have serum ferritin levels between 1000-2499 ng/mL, and 6 patients (33.33%) have serum ferritin levels within 2500-4999 ng/mL. Four patients (22.22) recorded serum ferritin levels between 5000-9999 ng/mL and only 1 patient (6.67%) has a serum ferritin level higher than 10,000 ng/mL.

Table 18.9: Distribution of Patients in Labuan According to Most Recent Serum Ferritin Level

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Labuan	18	5	27.78	2	11.11	6	33.33	4	22.22	1	6.67
Total	18	5	27.78	2	11.11	6	33.33	4	22.22	1	6.67

18.5 Conclusion

This report gives a brief overview of the number of thalassaemia patients in Labuan, the types of thalassaemia they have and the basic treatment available for these patients. Hospital Labuan handles only 27 patients with thalassaemia. The most common type of thalassaemia in Labuan is β-thalassaemia major.

Currently, the patients in Labuan are reviewed by Medical Officers in the thalassaemia clinic with the assistance of the visiting haematologist from Hospital Queen Elizabeth, Sabah every three months. Due to unavailability of a day care centre in this hospital, all thalassaemia patients receiving treatment must be admitted to ward and follow regular appointment in the clinic. A significant problem faced in Hospital Labuan is the limited number of nurses who are experienced in managing thalassaemia patients.

19 Wilayah Persekutuan Putrajaya

19.1 Introduction

Putrajaya is a planned city, located south of Kuala Lumpur. The city serves as the federal administrative centre of Malaysia. The seat of government was shifted from Kuala Lumpur to Putrajaya in 1999 due to overcrowding and congestion in Kuala Lumpur. Nevertheless, Kuala Lumpur remains as Malaysia's national capital and also as the commercial and financial centre. Putrajaya was the brainchild of former Prime Minister Tun Dr. Mahathir Mohamad. In 2001, Putrajaya was made into a Federal Territory, increasing the number of federal territories of Malaysia to three (after Kuala Lumpur and Labuan).

Putrajaya was named after the first Malaysian Prime Minister, Tunku Abdul Rahman Putra. The city is situated within the Multimedia Super Corridor, neighbouring the also newly-developed Cyberjaya. Putrajaya's development began in 1995; today, major landmarks have been completed and the population is expected to grow in the relatively new city. "Jaya" in the city's name translates to "victory" in Sanskrit. As of 2019, the population of Putrajaya is around 97,000.

19.2 Patient Demographics

Data were analysed from patients who are either living, lost to follow-up or cured by transplant, whereas deceased patients were excluded. In total, there are 36 living patients treated in Hospital Putrajaya (Table 19.1). Thirty-three patients are on active treatment and another 3 patients have undergone stem cell therapy (Table 19.2). Table 19.3 shows that there is only 1 thalassaemia patient death in Putrajaya since 2007, caused by infection.

Table 19.1: Distribution of Patients in Putrajaya by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Putrajaya	36	100.00
Total	36	100.00

Table 19.2: Distribution of Patients in Putrajaya by Vital Status

Status	Number of Patients
Alive and On Active Treatment	33
Cured by Stem Cell Therapy	3
Total	**36**
Lost to Follow-up	0
Total	**36**
Deaths in 2019	0
Cumulative Reported Deaths	1

Table 19.3: Cumulative Cause of Death in Putrajaya Since 1997

Cause of Death	Number of Patients
Infection	1
Total	1

19.2.1. Age Group

The youngest patient in Putrajaya is 3 years old and the eldest patient is 45 years old (diagnosed with β-thalassaemia intermedia). The age group with the highest number of patients in Putrajaya is 5-9.9 years with 13 patients (Figure 19.1). Most patients are diagnosed with HbE/β-thalassaemia, as shown in Table 19.4.

Figure 19.1: Distribution of Patients in Putrajaya by Age Group

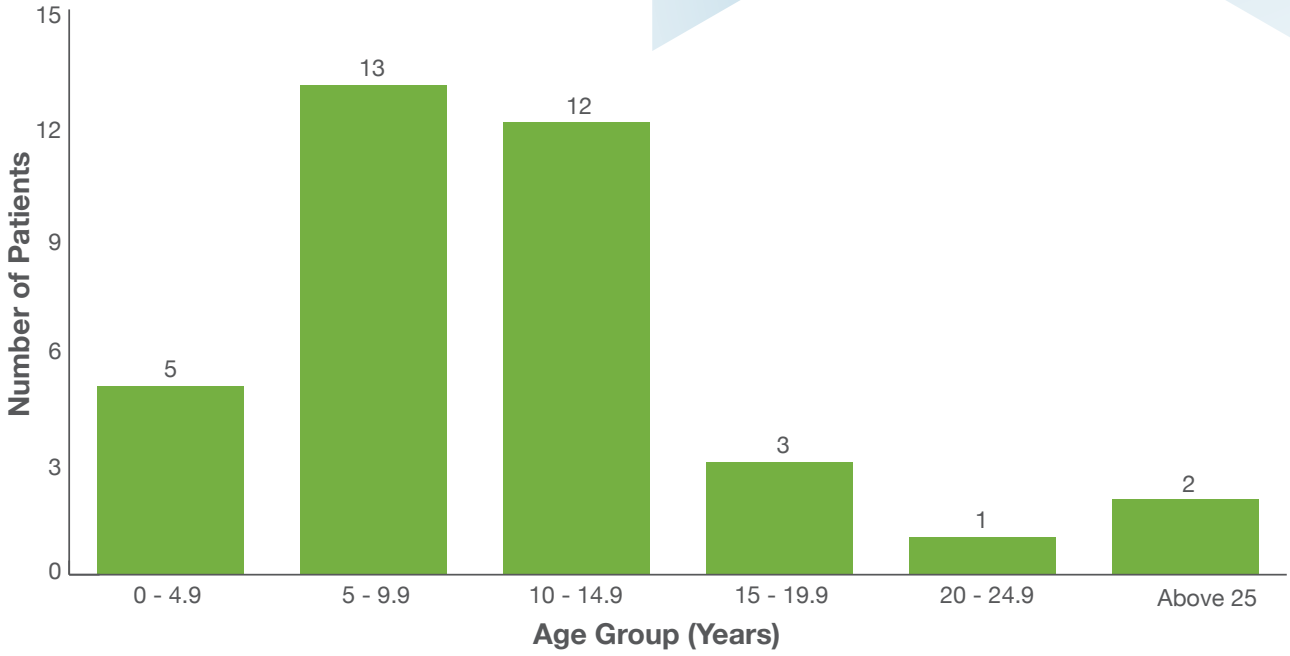


Table 19.4: Distribution of Patients in Putrajaya According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 14.9	30	β-Thalassaemia Major	7	23.33
		β-Thalassaemia Intermedia	3	10.00
		HbE/β-Thalassaemia	18	60.00
		HbH Disease	2	6.67
		Others	0	0.00
15 - 29.9	6	β-Thalassaemia Major	1	16.67
		β-Thalassaemia Intermedia	1	16.67
		HbE/β-Thalassaemia	4	66.67
		HbH Disease	0	0.00
		Others	0	0.00
Total			36	

19.2.2 Gender

Table 19.5 shows the distribution of patients in Putrajaya by gender. Thirty patients (83.33%) are male and 6 patients (16.67%) are female.

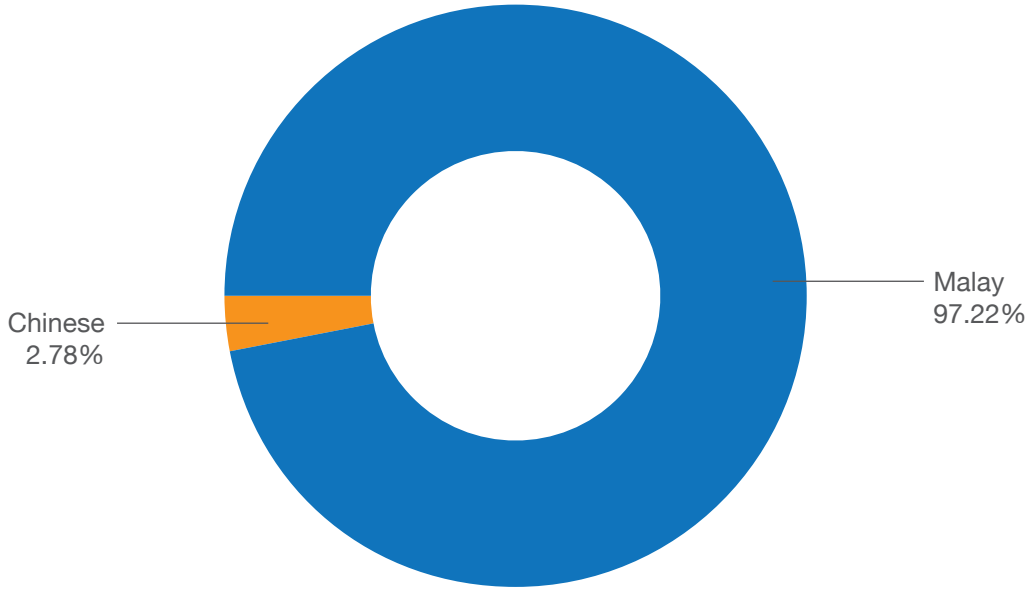
Table 19.5: Distribution of Patients in Putrajaya by Gender

Centre	Male		Female	
	No.	%	No.	%
Hospital Putrajaya	30	83.33	6	16.67
Total	30	83.33	6	16.67

19.2.3 Ethnic Group

Most patients in Putrajaya (35 patients, 97.22%) are Malay and 1 patient (2.78%) is Chinese (Figure 19.2).

Figure 19.2: Distribution of Patients in Putrajaya by Ethnic Group



19.3 Diagnosis

Figure 19.3 shows that just over half of thalassaemia patients in Putrajaya have HbE/β-thalassaemia (22 patients, 61.11%), followed by β-thalassaemia major with 8 patients (22.22%). HbE/β-thalassaemia is the most common thalassaemia diagnosis afflicting Malay patients in Putrajaya. The only Chinese patient in Putrajaya has β-thalassaemia major (Table 19.6).

Figure 19.3: Distribution of Patients in Putrajaya by Diagnosis

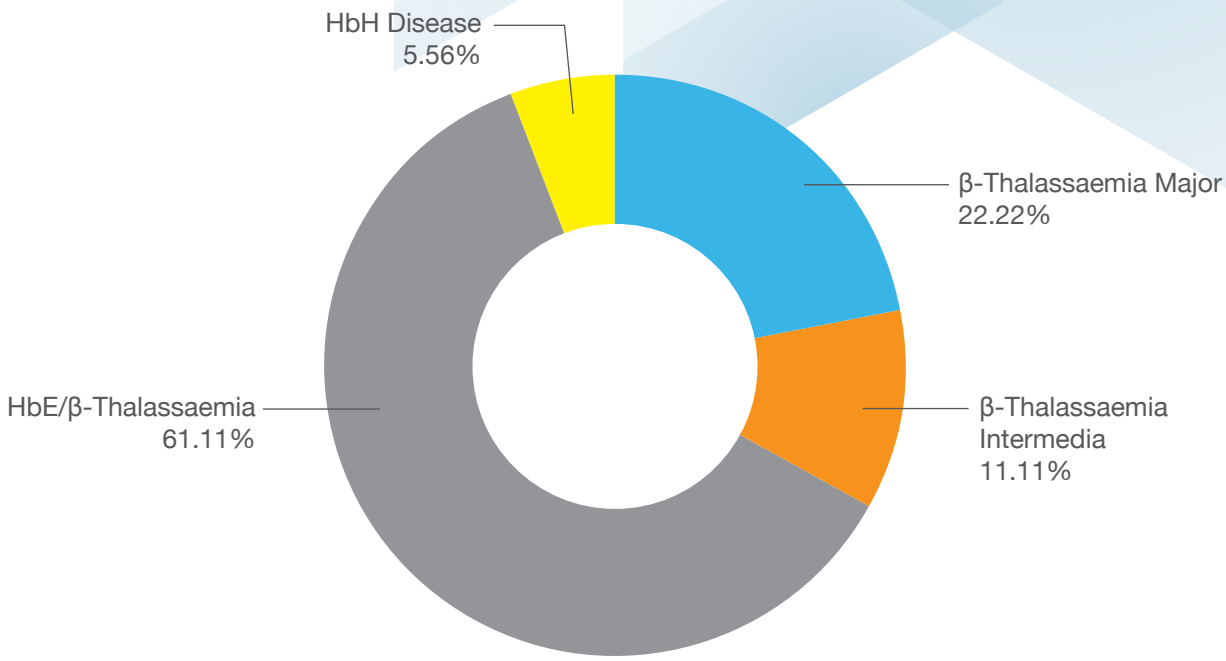


Table 19.6: Distribution of Patients in Putrajaya According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	8	Malay	7	19.44
		Chinese	1	2.78
β-Thalassaemia Intermedia	4	Malay	4	11.11
HbE/β-Thalassaemia	22	Malay	22	61.11
HbH Disease	2	Malay	2	5.56
Total			36	100.00

19.4 Treatment

19.4.1 Iron Chelation Therapy

As shown in Table 19.7, 23 patients in Putrajaya are receiving iron chelation therapy. Eighteen patients (78.26%) are on DFX, 4 patients (17.39%) are on DFO and 1 patient (4.35%) is on DFP monotherapy. The remaining 13 patients in Putrajaya are not on iron chelation therapy. Fifteen out of 18 patients receiving DFX therapy are below 15 years old (Table 19.8).

Table 19.7: Distribution of Patients in Putrajaya by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	4	17.39
DFP only	1	4.35
DFX only	18	78.26
DFO + DFP	0	0.00
DFP + DFX	0	0.00
DFO + DFX	0	0.00
DFO + DFP + DFX	0	0.00
Total	23	100.00

Table 19.8: Distribution of Patients in Putrajaya According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 14.9	19	DFO only	4	21.05
		DFP only	0	0.00
		DFX only	15	78.95
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15 - 29.9	4	DFO only	0	0.00
		DFP only	1	25.00
		DFX only	3	75.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			23	

19.4.2 Serum Ferritin Level

Twenty-two patients who receive regular transfusion have had their serum ferritin levels measured in 2019. Most of these patients have a serum ferritin level below 2499 ng/mL (17 patients, 77.27%), 4 patients (18.18%) have a serum ferritin level between 2500-4999 ng/mL, and 1 patient (4.55%) has a serum ferritin level above 5000 ng/mL (Table 19.9).

Table 19.9: Distribution of Patients in Putrajaya According to Most Recent Serum Ferritin Level

Centre	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Putrajaya	22	4	18.18	13	59.09	4	18.18	1	4.55	0	0
Total	22	4	18.18	13	59.09	4	18.18	1	4.55	0	0.00

19.5 Conclusion

There are 36 thalassaemia patients in Putrajaya. Thirty of these patients (83.33%) are male and 6 patients (16.67%) are female. Most patients in Putrajaya are of Malay descent (35 patients, 97.22%), and only 1 patient (2.78%) is of Chinese descent. Eighteen patients are children below 10 years of age.

The majority of patients in Putrajaya are diagnosed with HbE/β-thalassaemia (22 patients, 61.11%), followed by β-thalassaemia major (8 patients, 22.22%) and β-thalassaemia intermedia (4 patients, 11.11%). Twenty-three patients (61.11%) receive iron chelation therapy; 18 patients (78.26%) are on DFX monotherapy, followed by 4 patients (17.39%) on DFO monotherapy and 1 patient (4.35%) on DFP monotherapy.

Of the 36 patients in Putrajaya, 22 require regular transfusions. Based on serum ferritin levels recorded in 2019, most patients (17 out of 22 TDT patients, 77.27%) have a serum ferritin level below 2500 ng/mL. The remaining 5 patients have serum ferritin levels above 2500 ng/mL. Data on serum ferritin levels in Putrajaya were recorded only for TDT patients. The results show that the target serum ferritin level of below 2499 ng/mL has been achieved in more than 75% of the patients in Putrajaya.

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APPENDIX

APPENDIX A

Table A1: Number of Thalassaemia Patients in Malaysia by State, 2015 – 2019

State	2015		2016		2017		2018		2019	
	No.	%	No.	%	No.	%	No.	%	No.	%
Johor	541	7.50	587	7.72	617	7.83	637	7.98	643	7.86
Kedah	668	9.26	680	8.94	688	8.73	694	8.69	715	8.74
Kelantan	423	5.86	449	5.90	474	6.01	486	6.09	485	5.93
Melaka	199	2.76	209	2.75	220	2.79	226	2.83	246	3.01
Negeri Sembilan	172	2.38	180	2.37	180	2.28	181	2.27	202	2.47
Pahang	360	4.99	403	5.30	428	5.43	437	5.47	432	5.28
Perak	519	7.19	550	7.23	563	7.14	564	7.06	594	7.26
Perlis	120	1.66	124	1.63	127	1.61	128	1.60	126	1.54
Pulau Pinang	452	6.26	470	6.18	479	6.08	480	6.01	499	6.10
Sabah	1671	23.15	1740	22.88	1792	22.74	1814	22.72	1829	22.36
Sarawak	194	2.69	207	2.72	219	2.78	223	2.79	244	2.98
Selangor	1075	14.90	1124	14.78	1160	14.72	1169	14.64	1249	15.27
Terengganu	289	4.00	321	4.22	339	4.30	344	4.31	345	4.22
W. P. Kuala Lumpur	482	6.68	501	6.59	530	6.72	535	6.70	506	6.19
W. P. Labuan	19	0.26	20	0.26	25	0.32	25	0.31	27	0.33
W. P. Putrajaya	33	0.46	40	0.53	41	0.52	41	0.51	36	0.44
Total	7217	100.00	7605	100.00	7882	100.00	7984	100.00	8178	100.00

Table A2: Number of New Thalassaemia Births in Malaysia by State, 2012 – 2019

State	2012	2013	2014	2015	2016	2017	2018	2019
Johor	23	30	21	14	12	9	9	0
Kedah	26	16	14	8	6	3	2	0
Kelantan	14	13	8	11	7	3	1	0
Melaka	11	7	12	12	3	5	2	0
Negeri Sembilan	7	3	1	5	2	1	0	0
Pahang	10	16	14	10	5	4	1	0
Perak	14	12	9	15	4	5	2	0
Perlis	2	1	2	0	3	2	0	0
Pulau Pinang	13	12	9	7	3	4	0	1
Sabah	79	59	64	48	37	36	12	0
Sarawak	7	9	8	3	6	7	2	0
Selangor	27	32	28	17	16	15	3	1
Terengganu	8	10	10	12	11	5	3	0
W. P. Kuala Lumpur	15	17	19	10	10	15	2	1
W. P. Labuan	1	3	3	2	3	0	0	0
W. P. Putrajaya	0	3	4	3	2	0	0	0
Total	257	243	226	177	130	114	39	3

Table A3: Distribution of Patients in Malaysia According to Diagnosis by Year

Year	Total Number of Patients	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/ β-Thalassaemia		HbH Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
2019	8178	2671	32.66	738	9.02	2878	35.19	1593	19.48	298	3.64
2018	8098	2645	32.66	731	9.03	2852	35.22	1575	19.45	295	3.64
2017	7863	2591	32.95	711	9.04	2748	34.95	1530	19.46	324	4.12
2016	7539	2518	33.40	673	8.93	2628	34.86	1462	19.39	258	3.42
2015	7142	2427	33.98	651	9.12	2463	34.49	1371	19.20	228	3.19
2014	6707	2318	34.56	611	9.11	2324	34.65	1258	18.76	196	2.92
2013	6275	2217	35.33	574	9.15	2172	34.61	1138	18.14	174	2.77
2012	5844	2108	36.07	541	9.26	2021	34.58	1029	17.61	145	2.48
2011	5408	1997	36.93	501	9.26	1865	34.49	914	16.90	131	2.42
2010	5019	1907	38.00	462	9.21	1726	34.39	808	16.10	116	2.31
2009	4617	1781	38.57	422	9.14	1584	34.31	730	15.81	100	2.17

Table A4: Distribution of Patients in Malaysia According to Diagnosis by State

State	Total Number of Patients	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/ β-Thalassaemia		HbH Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Johor	643	199	30.95	42	6.53	291	45.26	109	16.95	2	0.31
Kedah	715	109	15.24	55	7.69	301	42.10	199	27.83	51	7.13
Kelantan	485	66	13.61	32	6.60	247	50.93	120	24.74	20	4.12
Melaka	246	56	22.76	9	3.66	102	41.46	65	26.42	14	5.69
Negeri Sembilan	202	54	26.73	22	10.89	76	37.62	31	15.35	19	9.41
Pahang	432	66	15.28	31	7.18	185	42.82	107	24.77	43	9.95
Perak	594	124	20.88	53	8.92	259	43.60	132	22.22	26	4.38
Perlis	126	27	21.43	16	12.70	54	42.86	24	19.05	5	3.97
Pulau Pinang	499	105	21.04	36	7.21	202	40.48	134	26.85	22	4.41
Sabah	1829	1321	72.23	274	14.98	130	7.11	99	5.41	5	0.27
Sarawak	244	101	41.39	17	6.97	52	21.31	74	30.33	0	0.00
Selangor	1249	222	17.77	92	7.37	547	43.80	333	26.66	55	4.40
Terengganu	345	62	17.97	30	8.70	184	53.33	63	18.26	6	1.74
W. P. Kuala Lumpur	506	136	26.88	16	3.16	226	44.66	100	19.76	28	5.53
W. P. Labuan	27	15	55.56	9	33.33	0	0.00	1	3.70	2	7.41
W. P. Putrajaya	36	8	22.22	4	11.11	22	61.11	2	5.56	0	0.00
Total	8178	2671	32.66	738	9.02	2878	35.19	1593	19.48	298	3.64

Table A5: Distribution of Patients in Malaysia According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/ β-Thalassaemia		HbH Disease		Other	
		No.	%	No.	%	No.	%	No.	%	No.	%
0 - 4.99	463	202	43.63	39	8.42	136	29.37	66	14.25	20	4.32
5 - 9.99	1264	425	33.62	103	8.15	419	33.15	244	19.30	73	5.78
10 - 14.9	1457	533	36.58	112	7.69	503	34.52	257	17.64	52	3.57
15 - 19.9	1276	463	36.29	111	8.70	475	37.23	195	15.28	32	2.51
20 - 24.9	1196	464	38.80	80	6.69	409	34.20	211	17.64	32	2.68
25 - 29.9	862	295	34.22	67	7.77	335	38.86	147	17.05	18	2.09
30 - 34.9	589	175	29.71	56	9.51	230	39.05	112	19.02	16	2.72
35 - 39.9	367	67	18.26	57	15.53	129	35.15	92	25.07	22	5.99
40 - 44.9	226	23	10.18	26	11.50	97	42.92	70	30.97	10	4.42
45 - 49.9	165	14	8.48	34	20.61	54	32.73	56	33.94	7	4.24
50 - 54.9	109	4	3.67	20	18.35	40	36.70	40	36.70	5	4.59
55 - 59.9	74	2	2.70	15	20.27	22	29.73	31	41.89	4	5.41
60 - 64.9	53	3	5.66	4	7.55	17	32.08	26	49.06	3	5.66
Above 65	77	1	1.30	14	18.18	12	15.58	46	59.74	4	5.19
Total	8178	2671	32.66	738	9.02	2878	35.19	1593	19.48	298	3.64

Table A6: Distribution of Patients in Malaysia According to Type of Iron Chelator Received by State

State	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Johor	428	DFO only	50	11.68
		DFP only	160	37.38
		DFX only	104	24.30
		DFO + DFP	90	21.03
		DFP + DFX	8	1.87
		DFO + DFX	15	3.50
		DFO + DFP + DFX	1	0.23
Kedah	391	DFO only	37	9.46
		DFP only	78	19.95
		DFX only	201	51.41
		DFO + DFP	48	12.28
		DFP + DFX	12	3.07
		DFO + DFX	14	3.58
		DFO + DFP + DFX	1	0.26
Kelantan	272	DFO only	46	16.91
		DFP only	60	22.06
		DFX only	126	46.32
		DFO + DFP	16	5.88
		DFP + DFX	9	3.31
		DFO + DFX	14	5.15
		DFO + DFP + DFX	1	0.37
Melaka	127	DFO only	33	25.98
		DFP only	17	13.39
		DFX only	53	41.73
		DFO + DFP	15	11.81
		DFP + DFX	2	1.57
		DFO + DFX	7	5.51
		DFO + DFP + DFX	0	0.00

Negeri Sembilan	135	DFO only	34	25.19
		DFP only	37	27.41
		DFX only	34	25.19
		DFO + DFP	24	17.78
		DFP + DFX	0	0.00
		DFO + DFX	6	4.44
		DFO + DFP + DFX	0	0.00
Pahang	253	DFO only	28	11.07
		DFP only	89	35.18
		DFX only	66	26.09
		DFO + DFP	53	20.95
		DFP + DFX	3	1.19
		DFO + DFX	13	5.14
		DFO + DFP + DFX	1	0.40
Perak	354	DFO only	81	22.88
		DFP only	72	20.34
		DFX only	128	36.16
		DFO + DFP	55	15.54
		DFP + DFX	8	2.26
		DFO + DFX	8	2.26
		DFO + DFP + DFX	2	0.56
Perlis	75	DFO only	3	4.00
		DFP only	3	4.00
		DFX only	64	85.33
		DFO + DFP	2	2.67
		DFP + DFX	1	1.33
		DFO + DFX	2	2.67
		DFO + DFP + DFX	0	0.00
Pulau Pinang	317	DFO only	58	18.30
		DFP only	98	30.91
		DFX only	77	24.29
		DFO + DFP	66	20.82
		DFP + DFX	6	1.89
		DFO + DFX	12	3.79
		DFO + DFP + DFX	0	0.00
Sabah	1228	DFO only	327	26.63
		DFP only	155	12.62
		DFX only	337	27.44
		DFO + DFP	270	21.99
		DFP + DFX	48	3.91
		DFO + DFX	60	4.89
		DFO + DFP + DFX	31	2.52
Sarawak	122	DFO only	10	8.20
		DFP only	21	17.21
		DFX only	53	43.44
		DFO + DFP	33	27.05
		DFP + DFX	1	0.82
		DFO + DFX	4	3.28
		DFO + DFP + DFX	0	0.00

Selangor	809	DFO only	74	9.15
		DFP only	328	40.54
		DFX only	169	20.89
		DFO + DFP	208	25.71
		DFP + DFX	7	0.87
		DFO + DFX	22	2.72
		DFO + DFP + DFX	1	0.12
Terengganu	239	DFO only	30	12.55
		DFP only	111	46.44
		DFX only	74	30.96
		DFO + DFP	24	10.04
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
W. P. Kuala Lumpur	293	DFO only	58	19.80
		DFP only	38	12.97
		DFX only	136	46.42
		DFO + DFP	48	16.38
		DFP + DFX	3	1.02
		DFO + DFX	10	3.41
		DFO + DFP + DFX	0	0.00
W. P. Labuan	18	DFO only	3	16.67
		DFP only	0	0.00
		DFX only	9	50.00
		DFO + DFP	3	16.67
		DFP + DFX	2	11.11
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	5.56
W. P. Putrajaya	23	DFO only	4	17.39
		DFP only	1	4.35
		DFX only	18	78.26
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			5084	100.00

Table A7: Distribution of Patients in Malaysia According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	144	DFO only	7	0.14
		DFP only	0	0.00
		DFX only	135	2.66
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	2	0.04
		DFO + DFP + DFX	0	0.00
5 - 9.9	706	DFO only	69	1.36
		DFP only	44	0.89
		DFX only	530	10.42
		DFO + DFP	13	0.26
		DFP + DFX	14	0.28
		DFO + DFX	34	0.67
		DFO + DFP + DFX	2	0.04
10 - 14.9	961	DFO only	108	2.12
		DFP only	152	2.99
		DFX only	541	10.64
		DFO + DFP	77	1.51
		DFP + DFX	32	0.63
		DFO + DFX	43	0.85
		DFO + DFP + DFX	8	0.16
15 - 19.9	881	DFO only	215	4.23
		DFP only	191	3.76
		DFX only	253	4.98
		DFO + DFP	172	3.38
		DFP + DFX	15	0.30
		DFO + DFX	33	0.65
		DFO + DFP + DFX	2	0.04
20 - 24.9	827	DFO only	209	4.11
		DFP only	173	3.40
		DFX only	101	1.99
		DFO + DFP	282	5.55
		DFP + DFX	19	0.37
		DFO + DFX	34	0.67
		DFO + DFP + DFX	9	0.18
25 - 29.9	571	DFO only	132	2.60
		DFP only	176	3.46
		DFX only	29	0.57
		DFO + DFP	188	3.70
		DFP + DFX	16	0.31
		DFO + DFX	19	0.37
		DFO + DFP + DFX	11	0.22
30 - 34.9	379	DFO only	66	1.30
		DFP only	155	3.05
		DFX only	25	0.49
		DFO + DFP	108	2.12
		DFP + DFX	7	0.14
		DFO + DFX	13	0.26
		DFO + DFP + DFX	5	0.10

35 - 39.9	212	DFO only	25	0.49
		DFP only	118	2.32
		DFX only	9	0.18
		DFO + DFP	54	1.06
		DFP + DFX	2	0.04
		DFO + DFX	2	0.04
		DFO + DFP + DFX	2	0.04
40 - 44.9	144	DFO only	12	0.24
		DFP only	97	1.91
		DFX only	7	0.14
		DFO + DFP	24	0.47
		DFP + DFX	2	0.04
		DFO + DFX	2	0.04
		DFO + DFP + DFX	0	0.00
45 - 49.9	98	DFO only	15	0.30
		DFP only	58	1.14
		DFX only	7	0.14
		DFO + DFP	14	0.28
		DFP + DFX	1	0.02
		DFO + DFX	3	0.06
		DFO + DFP + DFX	0	0.00
50 - 54.9	62	DFO only	9	0.18
		DFP only	39	0.77
		DFX only	3	0.06
		DFO + DFP	9	0.18
		DFP + DFX	1	0.02
		DFO + DFX	1	0.02
		DFO + DFP + DFX	0	0.00
55 - 59.9	41	DFO only	3	0.06
		DFP only	27	0.53
		DFX only	3	0.06
		DFO + DFP	8	0.16
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	30	DFO only	3	0.06
		DFP only	20	0.39
		DFX only	2	0.04
		DFO + DFP	4	0.08
		DFP + DFX	0	0.00
		DFO + DFX	1	0.02
		DFO + DFP + DFX	0	0.00
Above 65	28	DFO only	3	0.06
		DFP only	18	0.35
		DFX only	4	0.08
		DFO + DFP	2	0.04
		DFP + DFX	1	0.02
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			5084	100.00

Table A8: Distribution of Patients in Malaysia According to Most Recent Serum Ferritin Level by Year

Year	Total Number of Patients	Serum Ferritin Level (ng/mL)									
		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
2019	2912	532	18.27	1044	35.85	797	27.37	440	15.11	99	3.40
2018	3709	678	18.28	1261	34.00	999	26.93	614	16.55	157	4.23
2017	3399	679	19.98	1098	32.30	888	26.13	605	17.80	129	3.80
2016	2893	538	18.60	935	32.32	794	27.45	506	17.49	120	4.15
2015	2667	489	18.34	842	31.57	795	29.81	428	16.05	113	4.24
2014	2122	350	16.49	739	34.83	626	29.50	347	16.35	60	2.83
2013	2177	359	16.49	694	31.88	644	29.58	393	18.05	87	4.00
2012	2001	311	15.54	559	27.94	594	29.69	411	20.54	126	6.30
2011	1707	268	15.70	432	25.31	528	30.93	384	22.50	94	5.51
2010	2107	266	12.62	567	26.91	653	30.99	486	23.07	135	6.41
2009	1982	281	14.18	535	26.99	579	29.21	459	23.16	128	6.46
2008	1803	270	14.98	476	26.40	512	28.40	417	23.13	128	7.10
2007	1522	261	17.15	406	26.68	391	25.69	339	22.27	125	8.21

APPENDIX B

Table B1: Distribution of Patients in Johor According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	44	β-Thalassaemia Major	19	2.95
		β-Thalassaemia Intermedia	2	0.31
		HbE/β-Thalassaemia	17	2.64
		HbH Disease	6	0.93
		Others	0	0.00
5 - 9.9	109	β-Thalassaemia Major	30	4.67
		β-Thalassaemia Intermedia	3	0.47
		HbE/β-Thalassaemia	49	7.62
		HbH Disease	25	3.89
		Others	2	0.31
10 -14.9	104	β-Thalassaemia Major	23	3.58
		β-Thalassaemia Intermedia	6	0.93
		HbE/β-Thalassaemia	53	8.24
		HbH Disease	22	3.42
		Others	0	0.00
15 -19.9	106	β-Thalassaemia Major	38	5.91
		β-Thalassaemia Intermedia	3	0.47
		HbE/β-Thalassaemia	49	7.62
		HbH Disease	16	2.49
		Others	0	0.00
20 - 24.9	91	β-Thalassaemia Major	35	5.44
		β-Thalassaemia Intermedia	5	0.78
		HbE/β-Thalassaemia	38	5.91
		HbH Disease	13	2.02
		Others	0	0.00
25 - 29.9	64	β-Thalassaemia Major	25	3.89
		β-Thalassaemia Intermedia	1	0.16
		HbE/β-Thalassaemia	35	5.44
		HbH Disease	3	0.47
		Others	0	0.00
30 - 34.9	32	β-Thalassaemia Major	12	1.87
		β-Thalassaemia Intermedia	5	0.78
		HbE/β-Thalassaemia	12	1.87
		HbH Disease	3	0.47
		Others	0	0.00
35 - 39.9	35	β-Thalassaemia Major	8	1.24
		β-Thalassaemia Intermedia	7	1.09
		HbE/β-Thalassaemia	13	2.02
		HbH Disease	7	1.09
		Others	0	0.00
40 - 44.9	11	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	9	1.40
		HbH Disease	2	0.31
		Others	0	0.00

45 - 49.9	19	β-Thalassaemia Major	4	0.62
		β-Thalassaemia Intermedia	4	0.62
		HbE/β-Thalassaemia	7	1.09
		HbH Disease	4	0.62
		Others	0	0.00
50 - 54.9	10	β-Thalassaemia Major	2	0.31
		β-Thalassaemia Intermedia	2	0.31
		HbE/β-Thalassaemia	5	0.78
		HbH Disease	1	0.16
		Others	0	0.00
55 - 59.9	8	β-Thalassaemia Major	2	0.31
		β-Thalassaemia Intermedia	2	0.31
		HbE/β-Thalassaemia	3	0.47
		HbH Disease	1	0.16
		Others	0	0.00
60 - 64.9	8	β-Thalassaemia Major	1	0.16
		β-Thalassaemia Intermedia	2	0.31
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	5	0.78
		Others	0	0.00
Above 65	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.16
		HbH Disease	1	0.16
		Others	0	0.00
Total			643	100.00

Table B2: Distribution of Patients in Johor According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	13	DFO only	1	0.23
		DFP only	0	0.00
		DFX only	12	2.80
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	58	DFO only	4	0.93
		DFP only	4	0.93
		DFX only	44	10.28
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	6	1.40
		DFO + DFP + DFX	0	0.00
10 - 14.9	73	DFO only	6	1.40
		DFP only	28	6.54
		DFX only	32	7.48
		DFO + DFP	5	1.17
		DFP + DFX	1	0.23
		DFO + DFX	1	0.23
		DFO + DFP + DFX	0	0.00
15 - 19.9	75	DFO only	10	2.34
		DFP only	36	8.41
		DFX only	13	3.04
		DFO + DFP	11	2.57
		DFP + DFX	2	0.47
		DFO + DFX	3	0.70
		DFO + DFP + DFX	0	0.00
20 - 24.9	74	DFO only	13	3.04
		DFP only	23	5.37
		DFX only	2	0.47
		DFO + DFP	32	7.48
		DFP + DFX	2	0.47
		DFO + DFX	2	0.47
		DFO + DFP + DFX	0	0.00
25 - 29.9	42	DFO only	8	1.87
		DFP only	18	4.21
		DFX only	1	0.23
		DFO + DFP	13	3.04
		DFP + DFX	2	0.47
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 34.9	25	DFO only	3	0.70
		DFP only	6	1.40
		DFX only	0	0.00
		DFO + DFP	14	3.27
		DFP + DFX	0	0.00
		DFO + DFX	2	0.47
		DFO + DFP + DFX	0	0.00

35 - 39.9	26	DFO only	1	0.23
		DFP only	18	4.21
		DFX only	0	0.00
		DFO + DFP	4	0.93
		DFP + DFX	1	0.23
		DFO + DFX	1	0.23
		DFO + DFP + DFX	1	0.23
40 - 44.9	8	DFO only	0	0.00
		DFP only	6	1.40
		DFX only	0	0.00
		DFO + DFP	2	0.47
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	16	DFO only	2	0.47
		DFP only	11	2.57
		DFX only	0	0.00
		DFO + DFP	3	0.70
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	8	DFO only	2	0.47
		DFP only	3	0.70
		DFX only	0	0.00
		DFO + DFP	3	0.70
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	5	DFO only	0	0.00
		DFP only	3	0.70
		DFX only	0	0.00
		DFO + DFP	2	0.47
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	4	DFO only	0	0.00
		DFP only	3	0.70
		DFX only	0	0.00
		DFO + DFP	1	0.23
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	1	DFO only	0	0.00
		DFP only	1	0.23
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			428	100.00

APPENDIX C

Table C1: Distribution of Patients in Kedah According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 – 4.9	19	β-Thalassaemia Major	4	0.56
		β-Thalassaemia Intermedia	6	0.84
		HbE/β-Thalassaemia	6	0.84
		HbH Disease	3	0.42
		Others	0	0.00
5 – 9.9	118	β-Thalassaemia Major	15	2.10
		β-Thalassaemia Intermedia	10	1.44
		HbE/β-Thalassaemia	46	6.43
		HbH Disease	41	5.73
		Others	6	0.84
10 – 14.9	166	β-Thalassaemia Major	19	2.66
		β-Thalassaemia Intermedia	10	1.40
		HbE/β-Thalassaemia	73	10.21
		HbH Disease	48	6.71
		Others	16	2.24
15 – 19.9	140	β-Thalassaemia Major	21	2.94
		β-Thalassaemia Intermedia	7	0.98
		HbE/β-Thalassaemia	66	9.23
		HbH Disease	34	4.76
		Others	12	1.68
20 - 24.9	129	β-Thalassaemia Major	24	3.36
		β-Thalassaemia Intermedia	5	0.70
		HbE/β-Thalassaemia	52	7.27
		HbH Disease	39	5.45
		Others	9	1.23
25 – 29.9	77	β-Thalassaemia Major	14	1.96
		β-Thalassaemia Intermedia	6	0.84
		HbE/β-Thalassaemia	37	5.17
		HbH Disease	16	2.24
		Others	4	0.56
30 – 34.9	28	β-Thalassaemia Major	8	1.12
		β-Thalassaemia Intermedia	2	0.28
		HbE/β-Thalassaemia	11	1.54
		HbH Disease	7	0.98
		Others	0	0.00
35 – 39.9	15	β-Thalassaemia Major	1	0.14
		β-Thalassaemia Intermedia	4	0.56
		HbE/β-Thalassaemia	7	0.98
		HbH Disease	2	0.28
		Others	1	0.14
40 – 44.9	9	β-Thalassaemia Major	2	0.28
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	2	0.28
		HbH Disease	4	0.56
		Others	1	0.14

45 – 49.9	5	β-Thalassaemia Major	1	0.14
		β-Thalassaemia Intermedia	1	0.14
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	2	0.28
		Others	1	0.14
50 – 54.9	4	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	0.28
		HbE/β-Thalassaemia	1	0.14
		HbH Disease	1	0.14
		Others	0	0.00
55 - 59.9	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.14
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	0.14
		Others	0	0.00
60 - 64.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	3	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.14
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	0.14
		Others	1	0.14
Total			715	100.00

Table C2: Distribution of Patients in Kedah According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	11	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	11	2.81
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	56	DFO only	2	0.51
		DFP only	2	0.51
		DFX only	50	12.79
		DFO + DFP	0	0.00
		DFP + DFX	1	0.26
		DFO + DFX	1	0.26
		DFO + DFP + DFX	0	0.00
10 - 14.9	85	DFO only	2	0.51
		DFP only	9	2.30
		DFX only	68	17.39
		DFO + DFP	5	1.28
		DFP + DFX	0	0.00
		DFO + DFX	1	0.26
		DFO + DFP + DFX	0	0.00
15 -19.9	87	DFO only	8	2.05
		DFP only	12	3.07
		DFX only	49	12.53
		DFO + DFP	15	3.84
		DFP + DFX	1	0.26
		DFO + DFX	2	0.51
		DFO + DFP + DFX	0	0.00
20 - 24.9	66	DFO only	11	2.81
		DFP only	16	4.09
		DFX only	15	3.84
		DFO + DFP	12	3.07
		DFP + DFX	6	1.53
		DFO + DFX	6	1.53
		DFO + DFP + DFX	0	0.00
25 - 29.9	46	DFO only	13	3.32
		DFP only	18	4.60
		DFX only	2	0.51
		DFO + DFP	7	1.79
		DFP + DFX	3	0.77
		DFO + DFX	3	0.77
		DFO + DFP + DFX	0	0.00
30 - 34.9	21	DFO only	0	0.00
		DFP only	11	2.81
		DFX only	1	0.26
		DFO + DFP	7	1.79
		DFP + DFX	0	0.00
		DFO + DFX	2	0.51
		DFO + DFP + DFX	0	0.00

35 - 39.9	8	DFO only	0	0.00
		DFP only	6	1.53
		DFX only	0	0.00
		DFO + DFP	2	0.51
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	4	DFO only	0	0.00
		DFP only	2	0.51
		DFX only	2	0.51
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	2	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	0.26
		DFO + DFP	0	0.00
		DFP + DFX	1	0.26
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	2	DFO only	0	0.00
		DFP only	2	0.51
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	2	DFO only	1	0.26
		DFP only	0	0.00
		DFX only	1	0.26
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	0.26
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			391	100

APPENDIX D

Table D1: Distribution of Patients in Kelantan According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 – 4.9	22	β-Thalassaemia Major	6	1.24
		β-Thalassaemia Intermedia	2	0.41
		HbE/β-Thalassaemia	8	1.65
		HbH Disease	5	1.03
		Others	1	0.21
5 – 9.9	63	β-Thalassaemia Major	6	1.24
		β-Thalassaemia Intermedia	3	0.62
		HbE/β-Thalassaemia	33	6.80
		HbH Disease	17	3.51
		Others	4	0.82
10 – 14.9	89	β-Thalassaemia Major	14	2.89
		β-Thalassaemia Intermedia	9	1.86
		HbE/β-Thalassaemia	36	7.42
		HbH Disease	24	4.95
		Others	6	1.24
15 – 19.9	72	β-Thalassaemia Major	10	2.06
		β-Thalassaemia Intermedia	2	0.41
		HbE/β-Thalassaemia	44	9.07
		HbH Disease	16	3.30
		Others	0	0.00
20 - 24.9	89	β-Thalassaemia Major	7	1.44
		β-Thalassaemia Intermedia	1	0.21
		HbE/β-Thalassaemia	55	11.34
		HbH Disease	23	4.74
		Others	3	0.62
25 – 29.9	56	β-Thalassaemia Major	11	2.27
		β-Thalassaemia Intermedia	1	0.21
		HbE/β-Thalassaemia	27	5.57
		HbH Disease	16	3.30
		Others	1	0.21
30 – 34.9	40	β-Thalassaemia Major	7	1.44
		β-Thalassaemia Intermedia	3	0.62
		HbE/β-Thalassaemia	23	4.74
		HbH Disease	7	1.44
		Others	0	0.00
35 - 39.9	15	β-Thalassaemia Major	4	0.82
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	6	1.24
		HbH Disease	4	0.82
		Others	1	0.21
40 – 44.9	9	β-Thalassaemia Major	1	0.21
		β-Thalassaemia Intermedia	2	0.41
		HbE/β-Thalassaemia	5	1.03
		HbH Disease	1	0.21
		Others	0	0.00

45 - 49.9	15	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	5	1.03
		HbE/β-Thalassaemia	5	1.03
		HbH Disease	4	0.82
		Others	1	0.21
50 - 54.9	6	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.21
		HbE/β-Thalassaemia	2	0.41
		HbH Disease	1	0.21
		Others	2	0.41
55 - 59.9	3	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.21
		HbE/β-Thalassaemia	1	0.21
		HbH Disease	1	0.21
		Others	0	0.00
60 - 64.9	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.21
		HbE/β-Thalassaemia	1	0.21
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	4	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.21
		HbE/β-Thalassaemia	1	0.21
		HbH Disease	1	0.21
		Others	1	0.21
Total			485	100

Table D2: Distribution of Patients in Kelantan According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	4	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	4	1.47
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	31	DFO only	2	0.74
		DFP only	2	0.74
		DFX only	26	9.56
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.37
		DFO + DFP + DFX	0	0.00
10 - 14.9	55	DFO only	6	2.21
		DFP only	7	2.57
		DFX only	36	13.24
		DFO + DFP	1	0.37
		DFP + DFX	1	0.37
		DFO + DFX	4	1.47
		DFO + DFP + DFX	0	0.00
15 - 19.9	54	DFO only	10	3.68
		DFP only	14	5.15
		DFX only	26	9.56
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	4	1.47
		DFO + DFP + DFX	0	0.00
20 - 24.9	48	DFO only	13	4.78
		DFP only	8	2.94
		DFX only	18	6.62
		DFO + DFP	3	1.10
		DFP + DFX	2	0.74
		DFO + DFX	3	1.10
		DFO + DFP + DFX	1	0.37
25 - 29.9	31	DFO only	8	2.94
		DFP only	9	3.31
		DFX only	6	2.21
		DFO + DFP	5	1.84
		DFP + DFX	3	1.10
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 34.9	21	DFO only	2	0.74
		DFP only	8	2.94
		DFX only	6	2.21
		DFO + DFP	2	0.74
		DFP + DFX	1	0.37
		DFO + DFX	2	0.74
		DFO + DFP + DFX	0	0.00

35 - 39.9	7	DFO only	1	0.37
		DFP only	2	0.74
		DFX only	1	0.37
		DFO + DFP	2	0.74
		DFP + DFX	1	0.37
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	5	DFO only	2	0.74
		DFP only	2	0.74
		DFX only	1	0.37
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	9	DFO only	1	0.37
		DFP only	5	1.84
		DFX only	1	0.37
		DFO + DFP	2	0.74
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50-54.9	4	DFO only	0	0.00
		DFP only	1	0.37
		DFX only	1	0.37
		DFO + DFP	1	0.37
		DFP + DFX	1	0.37
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	1	DFO only	0	0.00
		DFP only	1	0.37
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	2	DFO only	1	0.37
		DFP only	1	0.37
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			272	100.00

APPENDIX E

Table E1: Distribution of Patients in Melaka According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 – 4.9	22	β-Thalassaemia Major	9	3.66
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	5	2.03
		HbH Disease	7	2.85
		Others	1	0.41
5 - 9.9	48	β-Thalassaemia Major	6	2.44
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	20	8.13
		HbH Disease	15	6.10
		Others	6	2.44
10 - 14.9	50	β-Thalassaemia Major	11	4.47
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	17	6.91
		HbH Disease	18	7.32
		Others	3	1.22
15 - 19.9	37	β-Thalassaemia Major	5	2.03
		β-Thalassaemia Intermedia	3	1.22
		HbE/β-Thalassaemia	22	8.94
		HbH Disease	5	2.03
		Others	2	0.81
20 - 24.9	38	β-Thalassaemia Major	10	4.07
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	16	6.50
		HbH Disease	11	4.47
		Others	1	0.41
25 - 29.9	28	β-Thalassaemia Major	10	4.07
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	14	5.69
		HbH Disease	2	0.81
		Others	1	0.41
30 - 34.9	17	β-Thalassaemia Major	4	1.63
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	6	2.44
		HbH Disease	6	2.44
		Others	0	0.00
35 - 39.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
40 – 44.9	2	β-Thalassaemia Major	1	0.41
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.41
		HbH Disease	0	0.00
		Others	0	0.00

45 - 49.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
50 - 54.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
55 - 59.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.41
		HbH Disease	0	0.00
		Others	0	0.00
60 - 64.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	0.41
		Others	0	0.00
Above 65	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			246	100.00

Table E2: Distribution of Patients in Melaka According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	4	DFO	0	0.00
		DFP	0	0.00
		DFX	4	3.15
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	16	DFO only	2	1.57
		DFP only	0	0.00
		DFX only	10	7.87
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	4	3.15
		DFO + DFP + DFX	0	0.00
10 - 14.9	25	DFO only	3	2.36
		DFP only	0	0.00
		DFX only	22	17.32
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15 - 19.9	20	DFO only	7	5.51
		DFP only	1	0.79
		DFX only	11	8.66
		DFO + DFP	1	0.79
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20 - 24.9	26	DFO only	8	6.30
		DFP only	4	3.15
		DFX only	7	5.51
		DFO + DFP	5	3.94
		DFP + DFX	1	0.79
		DFO + DFX	1	0.79
		DFO + DFP + DFX	0	0.00
25 - 29.9	18	DFO only	6	4.72
		DFP only	2	1.57
		DFX only	0	0.00
		DFO + DFP	9	7.09
		DFP + DFX	1	0.79
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 34.9	12	DFO	7	5.51
		DFP	3	2.36
		DFX	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	2	1.57
		DFO + DFP + DFX	0	0.00

35 - 39.9	1	DFO only	0	0.00
		DFP only	1	0.79
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	2	DFO only	0	0.00
		DFP only	2	1.57
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	1	DFO only	0	0.00
		DFP only	1	0.79
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	1	DFO	0	0.00
		DFP	1	0.79
		DFX	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	1	DFO only	0	0.00
		DFP only	1	0.79
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			127	100.00

APPENDIX F

Table F1: Distribution of Patients in Negeri Sembilan According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	8	β-Thalassaemia Major	2	0.99
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	4	1.98
		HbH Disease	1	0.50
		Others	1	0.50
5 - 9.9	25	β-Thalassaemia Major	9	4.46
		β-Thalassaemia Intermedia	2	0.99
		HbE/β-Thalassaemia	7	3.47
		HbH Disease	5	2.48
		Others	2	0.99
10 - 14.9	26	β-Thalassaemia Major	6	2.97
		β-Thalassaemia Intermedia	2	0.99
		HbE/β-Thalassaemia	12	5.94
		HbH Disease	2	0.99
		Others	4	1.98
15 - 19.9	29	β-Thalassaemia Major	9	4.46
		β-Thalassaemia Intermedia	1	0.50
		HbE/β-Thalassaemia	10	4.95
		HbH Disease	4	1.98
		Others	5	2.48
20 - 24.9	37	β-Thalassaemia Major	8	3.96
		β-Thalassaemia Intermedia	1	0.50
		HbE/β-Thalassaemia	21	10.40
		HbH Disease	5	2.48
		Others	2	0.99
25 - 29.9	24	β-Thalassaemia Major	11	5.45
		β-Thalassaemia Intermedia	1	0.50
		HbE/β-Thalassaemia	6	2.97
		HbH Disease	4	1.98
		Others	2	0.99
30 - 34.9	16	β-Thalassaemia Major	6	2.97
		β-Thalassaemia Intermedia	1	0.50
		HbE/β-Thalassaemia	5	2.48
		HbH Disease	3	1.49
		Others	1	0.50
35 - 39.9	9	β-Thalassaemia Major	1	0.50
		β-Thalassaemia Intermedia	4	1.98
		HbE/β-Thalassaemia	3	1.49
		HbH Disease	0	0.00
		Others	1	0.50
40 - 44.9	9	β-Thalassaemia Major	1	0.50
		β-Thalassaemia Intermedia	2	0.99
		HbE/β-Thalassaemia	3	1.49
		HbH Disease	2	0.99
		Others	1	0.50

45 - 49.9	4	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.50
		HbE/β-Thalassaemia	2	0.99
		HbH Disease	1	0.50
		Others	0	0.00
50 - 54.9	7	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	4	1.98
		HbE/β-Thalassaemia	1	0.50
		HbH Disease	2	0.99
		Others	0	0.00
55 - 59.9	4	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	3	1.49
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	0.50
		Others	0	0.00
60 - 64.9	3	β-Thalassaemia Major	1	0.50
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	2	0.99
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	0.50
		Others	0	0.00
Total			202	100.00

Table F2: Distribution of Patients in Negeri Sembilan According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	4	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	4	2.96
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP+ DFX	0	0.00
5 - 9.9	16	DFO only	4	2.96
		DFP only	1	0.74
		DFX only	10	7.41
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.74
		DFO + DFP+ DFX	0	0.00
10 -14.9	15	DFO only	1	0.74
		DFP only	0	0.00
		DFX only	13	9.63
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.74
		DFO + DFP + DFX	0	0.00
15 - 19.9	24	DFO only	12	8.89
		DFP only	2	1.48
		DFX only	3	2.22
		DFO + DFP	5	3.70
		DFP + DFX	0	0.00
		DFO + DFX	2	1.48
		DFO + DFP+ DFX	0	0.00
20 - 24.9	30	DFO only	8	5.93
		DFP only	6	4.44
		DFX only	2	1.48
		DFO + DFP	12	8.89
		DFP + DFX	0	0.00
		DFO + DFX	2	1.48
		DFO + DFP+ DFX	0	0.00
25 - 29.9	16	DFO only	3	2.22
		DFP only	9	6.67
		DFX only	0	0.00
		DFO + DFP	4	2.96
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP+ DFX	0	0.00
30 - 34.9	9	DFO only	2	1.48
		DFP only	5	3.70
		DFX only	1	0.74
		DFO + DFP	1	0.74
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP+ DFX	0	0.00

35 - 39.9	5	DFO only	2	1.48
		DFP only	3	2.22
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP+ DFX	0	0.00
40 - 44.9	6	DFO only	1	0.74
		DFP only	3	2.22
		DFX only	1	0.74
		DFO + DFP	1	0.74
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	2	DFO only	0	0.00
		DFP only	2	1.48
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP+ DFX	0	0.00
50 - 54.9	4	DFO only	0	0.00
		DFP only	4	2.96
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP+ DFX	0	0.00
55 - 59.9	2	DFO only	0	0.00
		DFP only	2	1.48
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP+ DFX	0	0.00
60 and above	2	DFO only	1	0.74
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	0.74
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP+ DFX	0	0.00
Total			135	100.00

APPENDIX G

Table G1: Distribution of Patients in Pahang According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	20	β-Thalassaemia Major	3	0.69
		β-Thalassaemia Intermedia	1	0.23
		HbE/β-Thalassaemia	9	2.08
		HbH Disease	4	0.93
		Others	3	0.69
5 - 9.9	74	β-Thalassaemia Major	7	1.62
		β-Thalassaemia Intermedia	6	1.39
		HbE/β-Thalassaemia	21	4.86
		HbH Disease	25	5.79
		Others	15	3.47
10 - 14.9	65	β-Thalassaemia Major	5	1.16
		β-Thalassaemia Intermedia	1	0.23
		HbE/β-Thalassaemia	37	8.56
		HbH Disease	20	4.63
		Others	2	0.46
15 -19.9	60	β-Thalassaemia Major	10	2.31
		β-Thalassaemia Intermedia	3	0.69
		HbE/β-Thalassaemia	31	7.18
		HbH Disease	14	3.24
		Others	2	0.46
20 - 24.9	50	β-Thalassaemia Major	13	3.01
		β-Thalassaemia Intermedia	4	0.93
		HbE/β-Thalassaemia	22	5.09
		HbH Disease	7	1.62
		Others	4	0.93
25 - 29.9	56	β-Thalassaemia Major	14	3.24
		β-Thalassaemia Intermedia	5	1.16
		HbE/β-Thalassaemia	22	5.09
		HbH Disease	11	2.55
		Others	4	0.93
30 - 34.9	37	β-Thalassaemia Major	5	1.16
		β-Thalassaemia Intermedia	5	1.16
		HbE/β-Thalassaemia	21	4.86
		HbH Disease	5	1.16
		Others	1	0.23
35 - 39.9	19	β-Thalassaemia Major	2	0.46
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	6	1.39
		HbH Disease	5	1.16
		Others	6	1.39
40 - 44.9	19	β-Thalassaemia Major	4	0.93
		β-Thalassaemia Intermedia	1	0.23
		HbE/β-Thalassaemia	8	1.85
		HbH Disease	5	1.16
		Others	1	0.23

45 - 49.9	10	β-Thalassaemia Major	3	0.69
		β-Thalassaemia Intermedia	1	0.23
		HbE/β-Thalassaemia	1	0.23
		HbH Disease	3	0.69
		Others	2	0.46
50 - 54.9	6	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	0.46
		HbE/β-Thalassaemia	2	0.46
		HbH Disease	1	0.23
		Others	1	0.23
55 - 59.9	9	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	0.46
		HbE/β-Thalassaemia	3	0.69
		HbH Disease	3	0.69
		Others	1	0.23
60 - 64.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	0.23
		Others	0	0.00
Above 65	6	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	2	0.46
		HbH Disease	3	0.69
		Others	1	0.23
Total			432	100.00

Table G2: Distribution of Patients in Pahang According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	20	DFO only	1	0.40
		DFP only	1	0.40
		DFX only	17	6.72
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.40
		DFO + DFP+ DFX	0	0.00
10 - 14.9	33	DFO only	1	0.40
		DFP only	5	1.98
		DFX only	26	10.28
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.40
		DFO + DFP + DFX	0	0.00
15 - 19.9	42	DFO only	5	1.98
		DFP only	7	2.77
		DFX only	17	6.72
		DFO + DFP	12	4.74
		DFP + DFX	0	0.00
		DFO + DFX	1	0.40
		DFO + DFP + DFX	0	0.00
20 - 24.9	41	DFO only	6	2.37
		DFP only	13	5.14
		DFX only	1	0.40
		DFO + DFP	15	5.93
		DFP + DFX	1	0.40
		DFO + DFX	4	1.58
		DFO + DFP + DFX	1	0.40
25 - 29.9	39	DFO only	6	2.37
		DFP only	20	7.91
		DFX only	3	1.19
		DFO + DFP	6	2.37
		DFP + DFX	0	0.00
		DFO + DFX	4	1.58
		DFO + DFP + DFX	0	0.00
30 - 34.9	25	DFO only	4	1.58
		DFP only	7	2.77
		DFX only	2	0.79
		DFO + DFP	9	3.56
		DFP + DFX	1	0.40
		DFO + DFX	2	0.79
		DFO + DFP+ DFX	0	0.00

35 - 39.9	16	DFO only	2	0.79
		DFP only	12	4.74
		DFX only	0	0.00
		DFO + DFP	2	0.79
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	15	DFO only	1	0.40
		DFP only	7	2.77
		DFX only	0	0.00
		DFO + DFP	6	2.37
		DFP + DFX	1	0.40
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	8	DFO only	1	0.40
		DFP only	6	2.37
		DFX only	0	0.00
		DFO + DFP	1	0.40
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	4	DFO only	0	0.00
		DFP only	3	1.19
		DFX only	0	0.00
		DFO + DFP	1	0.40
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	6	DFO only	1	0.40
		DFP only	4	1.58
		DFX only	0	0.00
		DFO + DFP	1	0.40
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	1	DFO only	0	0.00
		DFP only	1	0.40
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	3	DFO only	0	0.00
		DFP only	3	1.19
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			253	100.00

APPENDIX H

Table H1: Distribution of Patients in Perak According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	26	β-Thalassaemia Major	8	1.35
		β-Thalassaemia Intermedia	1	0.17
		HbE/β-Thalassaemia	13	2.19
		HbH Disease	3	0.51
		Others	1	0.17
5 - 9.9	66	β-Thalassaemia Major	12	2.02
		β-Thalassaemia Intermedia	3	0.51
		HbE/β-Thalassaemia	35	5.89
		HbH Disease	11	1.85
		Others	5	0.84
10 - 14.9	92	β-Thalassaemia Major	21	3.54
		β-Thalassaemia Intermedia	10	1.68
		HbE/β-Thalassaemia	39	6.57
		HbH Disease	20	3.37
		Others	2	0.34
15 - 19.9	76	β-Thalassaemia Major	17	2.86
		β-Thalassaemia Intermedia	10	1.68
		HbE/β-Thalassaemia	31	5.22
		HbH Disease	16	2.69
		Others	2	0.34
20 - 24.9	91	β-Thalassaemia Major	26	4.38
		β-Thalassaemia Intermedia	9	1.52
		HbE/β-Thalassaemia	42	7.07
		HbH Disease	13	2.19
		Others	1	0.17
25 - 29.9	74	β-Thalassaemia Major	14	2.36
		β-Thalassaemia Intermedia	5	0.84
		HbE/β-Thalassaemia	34	5.72
		HbH Disease	20	3.37
		Others	1	0.17
30 - 34.9	59	β-Thalassaemia Major	17	2.86
		β-Thalassaemia Intermedia	3	0.51
		HbE/β-Thalassaemia	29	4.88
		HbH Disease	7	1.18
		Others	3	0.51
35 - 39.9	35	β-Thalassaemia Major	8	1.35
		β-Thalassaemia Intermedia	4	0.67
		HbE/β-Thalassaemia	9	1.52
		HbH Disease	9	1.52
		Others	5	0.84
40 - 44.9	19	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	0.34
		HbE/β-Thalassaemia	5	0.84
		HbH Disease	10	1.68
		Others	2	0.34

45 - 49.9	18	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	10	1.68
		HbH Disease	7	1.18
		Others	1	0.17
50 - 54.9	12	β-Thalassaemia Major	1	0.17
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	5	0.84
		HbH Disease	5	0.84
		Others	1	0.17
55 - 59.9	10	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	3	0.51
		HbE/β-Thalassaemia	3	0.51
		HbH Disease	4	0.67
		Others	0	0.00
60 - 64.9	10	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	3	0.51
		HbH Disease	5	0.84
		Others	2	0.34
Above 65	6	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	3	0.51
		HbE/β-Thalassaemia	1	0.17
		HbH Disease	2	0.34
		Others	0	0.00
Total			594	100.00

Table H2: Distribution of Patients in Perak According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	0.28
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	36	DFO only	5	1.41
		DFP only	0	0.00
		DFX only	30	8.47
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.28
		DFO + DFP + DFX	0	0.00
10–14.9	59	DFO only	10	2.82
		DFP only	0	0.00
		DFX only	48	13.56
		DFO + DFP	1	0.28
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15 - 19.9	39	DFO only	11	3.11
		DFP only	4	1.13
		DFX only	22	6.21
		DFO + DFP	2	0.56
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20 - 24.9	66	DFO only	18	5.08
		DFP only	11	3.11
		DFX only	13	3.67
		DFO + DFP	18	5.08
		DFP + DFX	2	0.56
		DFO + DFX	3	0.85
		DFO + DFP + DFX	1	0.28
25 - 29.9	48	DFO only	14	3.95
		DFP only	7	1.98
		DFX only	4	1.13
		DFO + DFP	15	4.24
		DFP + DFX	4	1.13
		DFO + DFX	3	0.85
		DFO + DFP + DFX	1	0.28
30 - 34.9	40	DFO only	9	2.54
		DFP only	17	4.80
		DFX only	6	1.69
		DFO + DFP	7	1.98
		DFP + DFX	1	0.28
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

35–39.9	25	DFO only	5	1.41
		DFP only	11	3.11
		DFX only	1	0.28
		DFO + DFP	8	2.26
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	10	DFO only	4	1.13
		DFP only	5	1.41
		DFX only	1	0.28
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	9	DFO only	2	0.56
		DFP only	5	1.41
		DFX only	2	0.56
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	3	DFO only	0	0.00
		DFP only	2	0.56
		DFX only	0	0.00
		DFO + DFP	1	0.28
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	5	DFO only	1	0.28
		DFP only	3	0.85
		DFX only	0	0.00
		DFO + DFP	1	0.28
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60–64.9	9	DFO only	1	0.28
		DFP only	6	1.69
		DFX only	0	0.00
		DFO + DFP	1	0.28
		DFP + DFX	0	0.00
		DFO + DFX	1	0.28
		DFO + DFP + DFX	0	0.00
Above 65	4	DFO only	1	0.28
		DFP only	1	0.28
		DFX only	0	0.00
		DFO + DFP	1	0.28
		DFP + DFX	1	0.28
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			354	100.00

APPENDIX I

Table I1: Distribution of Patients in Perlis According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	5	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	3	2.38
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	2	1.59
		Others	0	0.00
5 - 9.9	10	β-Thalassaemia Major	1	0.79
		β-Thalassaemia Intermedia	3	2.38
		HbE/β-Thalassaemia	4	3.17
		HbH Disease	1	0.79
		Others	1	0.79
10 - 14.9	26	β-Thalassaemia Major	9	7.14
		β-Thalassaemia Intermedia	4	3.17
		HbE/β-Thalassaemia	6	4.76
		HbH Disease	6	4.76
		Others	1	0.79
15 - 19.9	21	β-Thalassaemia Major	3	2.38
		β-Thalassaemia Intermedia	4	3.17
		HbE/β-Thalassaemia	10	7.94
		HbH Disease	4	3.17
		Others	0	0.00
20 - 24.9	31	β-Thalassaemia Major	7	5.56
		β-Thalassaemia Intermedia	1	0.79
		HbE/β-Thalassaemia	13	10.32
		HbH Disease	9	7.14
		Others	1	0.79
25 - 29.9	19	β-Thalassaemia Major	3	2.38
		β-Thalassaemia Intermedia	1	0.79
		HbE/β-Thalassaemia	13	10.32
		HbH Disease	2	1.59
		Others	0	0.00
30 - 34.9	8	β-Thalassaemia Major	3	2.38
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	5	3.97
		HbH Disease	0	0.00
		Others	0	0.00
35 - 39.9	1	β-Thalassaemia Major	1	0.79
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
40 - 44.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.79
		HbH Disease	0	0.00
		Others	0	0.00

45 – 49.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.80
		HbH Disease	0	0.00
		Others	0	0.00
50 - 54.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
55 - 59.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	1	0.79
60 and above	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.79
		HbH Disease	0	0.00
		Others	1	0.79
Total			126	100.00

Table I2: Distribution of Patients in Perlis According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	3	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	3	4.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10–14.9	15	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	14	18.67
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	1.33
		DFO + DFP + DFX	0	0.00
15 - 19.9	17	DFO only	1	1.33
		DFP only	0	0.00
		DFX only	16	21.33
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20 - 24.9	19	DFO only	1	1.33
		DFP only	0	0.00
		DFX only	17	22.67
		DFO + DFP	1	1.33
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
25 - 29.9	12	DFO only	1	1.33
		DFP only	1	1.33
		DFX only	8	10.67
		DFO + DFP	1	1.33
		DFP + DFX	1	1.33
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 34.9	6	DFO only	0	0.00
		DFP only	1	1.33
		DFX only	4	5.33
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	1.33
		DFO + DFP + DFX	0	0.00

35 - 39.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	1.33
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	1.33
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	1	DFO only	0	0.00
		DFP only	1	1.33
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			75	100.00

APPENDIX J

Table J1: Distribution of Patients in Pulau Pinang According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 – 4.9	15	β-Thalassaemia Major	3	0.60
		β-Thalassaemia Intermedia	3	0.60
		HbE/β-Thalassaemia	4	0.80
		HbH Disease	5	1.00
		Others	0	0.00
5 - 9.9	55	β-Thalassaemia Major	9	1.80
		β-Thalassaemia Intermedia	4	0.80
		HbE/β-Thalassaemia	22	4.41
		HbH Disease	17	3.41
		Others	3	0.60
10 - 14.9	73	β-Thalassaemia Major	18	3.61
		β-Thalassaemia Intermedia	4	0.80
		HbE/β-Thalassaemia	35	7.01
		HbH Disease	12	2.40
		Others	4	0.80
15 - 19.9	74	β-Thalassaemia Major	18	3.61
		β-Thalassaemia Intermedia	4	0.80
		HbE/β-Thalassaemia	33	6.61
		HbH Disease	14	2.81
		Others	5	1.00
20 - 24.9	78	β-Thalassaemia Major	19	3.81
		β-Thalassaemia Intermedia	5	1.00
		HbE/β-Thalassaemia	31	6.21
		HbH Disease	21	4.21
		Others	2	0.40
25 - 29.9	53	β-Thalassaemia Major	20	4.01
		β-Thalassaemia Intermedia	1	0.20
		HbE/β-Thalassaemia	21	4.21
		HbH Disease	11	2.20
		Others	0	0.00
30 - 34.9	43	β-Thalassaemia Major	11	2.20
		β-Thalassaemia Intermedia	3	0.60
		HbE/β-Thalassaemia	18	3.61
		HbH Disease	6	1.20
		Others	5	1.00
35 - 39.9	31	β-Thalassaemia Major	6	1.20
		β-Thalassaemia Intermedia	5	1.00
		HbE/β-Thalassaemia	10	2.00
		HbH Disease	9	1.80
		Others	1	0.20
40 - 44.9	24	β-Thalassaemia Major	1	0.20
		β-Thalassaemia Intermedia	3	0.60
		HbE/β-Thalassaemia	13	2.61
		HbH Disease	6	1.20
		Others	1	0.20

45 - 49.9	18	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	0.40
		HbE/β-Thalassaemia	4	0.80
		HbH Disease	11	2.20
		Others	1	0.20
50 - 54.9	12	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.20
		HbE/β-Thalassaemia	4	0.80
		HbH Disease	7	1.40
		Others	0	0.00
55 - 59.9	12	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.20
		HbE/β-Thalassaemia	4	0.80
		HbH Disease	7	1.40
		Others	0	0.00
60 and above	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	2	0.40
		Others	0	0.00
Above 65	9	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	3	0.60
		HbH Disease	6	1.20
		Others	0	0.00
Total			499	100.00

Table J2: Distribution of Patients in Pulau Pinang According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	6	DFO only	1	0.32
		DFP only	0	0.00
		DFX only	5	1.58
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	29	DFO only	3	0.95
		DFP only	7	2.21
		DFX only	18	5.68
		DFO + DFP	1	0.32
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10 - 14.9	50	DFO only	11	3.47
		DFP only	7	2.21
		DFX only	18	5.68
		DFO + DFP	8	2.52
		DFP + DFX	2	0.63
		DFO + DFX	4	1.26
		DFO + DFP + DFX	0	0.00
15 - 19.9	50	DFO only	5	1.58
		DFP only	13	4.10
		DFX only	19	5.99
		DFO + DFP	10	3.15
		DFP + DFX	2	0.63
		DFO + DFX	1	0.32
		DFO + DFP + DFX	0	0.00
20 - 24.9	53	DFO only	7	2.21
		DFP only	14	4.42
		DFX only	12	3.79
		DFO + DFP	17	5.36
		DFP + DFX	1	0.32
		DFO + DFX	2	0.63
		DFO + DFP + DFX	0	0.00
25 - 29.9	35	DFO only	10	3.15
		DFP only	10	3.15
		DFX only	1	0.32
		DFO + DFP	12	3.79
		DFP + DFX	0	0.00
		DFO + DFX	2	0.63
		DFO + DFP + DFX	0	0.00
30 - 34.9	27	DFO only	5	1.58
		DFP only	12	3.79
		DFX only	2	0.63
		DFO + DFP	7	2.21
		DFP + DFX	0	0.00
		DFO + DFX	1	0.32
		DFO + DFP + DFX	0	0.00

35 - 39.9	21	DFO only	3	0.95
		DFP only	12	3.79
		DFX only	0	0.00
		DFO + DFP	6	1.89
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	17	DFO only	3	0.95
		DFP only	12	3.79
		DFX only	0	0.00
		DFO + DFP	1	0.32
		DFP + DFX	1	0.32
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	13	DFO only	5	1.58
		DFP only	5	1.58
		DFX only	0	0.00
		DFO + DFP	2	0.63
		DFP + DFX	0	0.00
		DFO + DFX	1	0.32
		DFO + DFP + DFX	0	0.00
50 - 54.9	9	DFO only	2	0.63
		DFP only	4	1.26
		DFX only	1	0.32
		DFO + DFP	1	0.32
		DFP + DFX	0	0.00
		DFO + DFX	1	0.32
		DFO + DFP + DFX	0	0.00
55 - 59.9	5	DFO only	1	0.32
		DFP only	2	0.63
		DFX only	1	0.32
		DFO + DFP	1	0.32
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	1	DFO only	1	0.32
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	1	DFO only	1	0.32
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			317	100.00

APPENDIX K

Table K1: Distribution of Patients in Sabah According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	133	β-Thalassaemia Major	104	5.69
		β-Thalassaemia Intermedia	9	0.49
		HbE/β-Thalassaemia	11	0.60
		HbH Disease	6	0.33
		Others	3	0.16
5 - 9.9	354	β-Thalassaemia Major	263	14.38
		β-Thalassaemia Intermedia	48	2.62
		HbE/β-Thalassaemia	28	1.53
		HbH Disease	15	0.82
		Others	0	0.00
10 -14.9	407	β-Thalassaemia Major	317	17.33
		β-Thalassaemia Intermedia	49	2.68
		HbE/β-Thalassaemia	26	1.42
		HbH Disease	14	0.77
		Others	1	0.50
15 -19.9	337	β-Thalassaemia Major	257	14.05
		β-Thalassaemia Intermedia	56	3.06
		HbE/β-Thalassaemia	14	0.77
		HbH Disease	10	0.55
		Others	0	0.00
20 - 24.9	252	β-Thalassaemia Major	212	11.59
		β-Thalassaemia Intermedia	31	1.69
		HbE/β-Thalassaemia	2	0.11
		HbH Disease	7	0.38
		Others	0	0.00
25 - 29.9	155	β-Thalassaemia Major	106	5.80
		β-Thalassaemia Intermedia	29	1.59
		HbE/β-Thalassaemia	16	0.87
		HbH Disease	4	0.22
		Others	0	0.00
30 - 34.9	78	β-Thalassaemia Major	41	2.24
		β-Thalassaemia Intermedia	18	0.98
		HbE/β-Thalassaemia	7	0.38
		HbH Disease	11	0.60
		Others	1	0.05
35 - 39.9	48	β-Thalassaemia Major	14	0.77
		β-Thalassaemia Intermedia	20	1.09
		HbE/β-Thalassaemia	7	0.38
		HbH Disease	7	0.38
		Others	0	0.00
40 - 44.9	22	β-Thalassaemia Major	5	0.27
		β-Thalassaemia Intermedia	7	0.38
		HbE/β-Thalassaemia	5	0.27
		HbH Disease	5	0.27
		Others	0	0.00

45 - 49.9	11	β-Thalassaemia Major	2	0.11
		β-Thalassaemia Intermedia	3	0.16
		HbE/β-Thalassaemia	1	0.05
		HbH Disease	5	0.27
		Others	0	0.00
50 - 54.9	13	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	0.11
		HbE/β-Thalassaemia	9	0.49
		HbH Disease	2	0.11
		Others	0	0.00
55 - 59.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.10
		HbH Disease	0	0.00
		Others	0	0.00
60 - 64.9	4	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.05
		HbH Disease	3	0.16
		Others	0	0.00
Above 65	14	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	0.11
		HbE/β-Thalassaemia	2	0.11
		HbH Disease	10	0.55
		Others	0	0.00
Total			1829	100.00

Table K2: Distribution of Patients in Sabah According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	34	DFO only	2	0.16
		DFP only	0	0.00
		DFX only	32	2.61
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	224	DFO only	21	1.71
		DFP only	8	0.65
		DFX only	165	13.44
		DFO + DFP	4	0.33
		DFP + DFX	10	0.81
		DFO + DFX	15	1.22
		DFO + DFP + DFX	1	0.08
10 - 14.9	300	DFO only	41	3.34
		DFP only	54	4.40
		DFX only	113	9.20
		Desferri oxamine + DFP	33	2.69
		DFP + DFX	28	2.28
		Deferri-oxamine + DFX	23	1.87
		DFO + DFP + DFX	8	0.65
15 - 19.9	253	DFO only	105	8.55
		DFP only	38	3.09
		DFX only	23	1.87
		DFO + DFP	67	5.46
		DFP + DFX	7	0.57
		DFO + DFX	12	0.98
		DFO + DFP + DFX	1	0.08
20 - 24.9	210	DFO only	99	8.06
		DFP only	10	0.81
		DFX only	4	0.33
		DFO + DFP	84	6.84
		DFP + DFX	1	0.08
		DFO + DFX	6	0.49
		DFO + DFP + DFX	6	0.49
25 - 29.9	119	DFO only	36	2.93
		DFP only	12	0.98
		DFX only	0	0.00
		DFO + DFP	57	4.64
		DFP + DFX	2	0.16
		DFO + DFX	2	0.16
		DFO + DFP + DFX	10	0.81
30 - 34.9	48	DFO only	13	1.06
		DFP only	15	1.22
		DFX only	0	0.00
		DFO + DFP	14	1.14
		DFP + DFX	0	0.00
		DFO + DFX	2	0.16
		DFO + DFP + DFX	4	0.33

35 - 39.9	23	DFO only	6	0.49
		DFP only	7	0.57
		DFX only	0	0.00
		DFO + DFP	9	0.73
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	0.08
40 - 44.9	9	DFO only	2	0.16
		DFP only	5	0.41
		DFX only	0	0.00
		DFO + DFP	2	0.16
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	4	DFO only	1	0.08
		DFP only	3	0.24
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	3	DFO only	1	0.08
		DFP only	2	0.16
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 and above	1	DFO only	0	0.00
		DFP only	1	0.08
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			1228	100.00

APPENDIX L

Table L1: Distribution of Patients in Sarawak According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 – 4.9	18	β-Thalassaemia Major	6	2.46
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	6	2.46
		HbH Disease	5	2.05
		Others	0	0.00
5 – 9.9	37	β-Thalassaemia Major	12	4.92
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	12	4.92
		HbH Disease	13	5.33
		Others	0	0.00
10 – 14.9	39	β-Thalassaemia Major	20	8.20
		β-Thalassaemia Intermedia	3	1.23
		HbE/β-Thalassaemia	8	3.28
		HbH Disease	8	3.28
		Others	0	0.00
15 – 19.9	32	β-Thalassaemia Major	16	6.56
		β-Thalassaemia Intermedia	2	0.82
		HbE/β-Thalassaemia	6	2.46
		HbH Disease	8	3.28
		Others	0	0.00
20 - 24.9	36	β-Thalassaemia Major	19	7.79
		β-Thalassaemia Intermedia	4	1.64
		HbE/β-Thalassaemia	6	2.46
		HbH Disease	7	2.87
		Others	0	0.00
25 – 29.9	24	β-Thalassaemia Major	13	5.33
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	5	2.05
		HbH Disease	5	2.05
		Others	0	0.00
30 – 34.9	25	β-Thalassaemia Major	12	4.92
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	12	4.92
		Others	0	0.00
35 – 39.9	14	β-Thalassaemia Major	3	1.23
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	4	1.64
		HbH Disease	6	2.46
		Others	0	0.00
40 – 44.5	3	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	2	0.82
		HbH Disease	0	0.00
		Others	0	0.00

45 – 49.9	5	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	2	0.82
		HbH Disease	2	0.82
		Others	0	0.00
50 – 54.9	5	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	1	0.41
		HbH Disease	3	1.23
		Others	0	0.00
55 - 59.9	2	β-Thalassaemia Major	0	0.0
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	2	0.82
		Others	0	0.00
60 – 64.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	4	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.41
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	3	1.23
		Others	0	0.00
Total			244	100.00

Table L2: Distribution of Patients in Sarawak According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	5	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	5	4.10
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
5-9.9	16	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	15	12.30
		DFO + DFP	1	0.82
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
10 - 14.9	25	DFO only	2	1.64
		DFP only	2	1.64
		DFX only	20	16.39
		DFO + DFP	1	0.82
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
15 – 19.9	18	DFO only	4	3.28
		DFP only	2	1.64
		DFX only	7	5.74
		DFO + DFP	5	4.10
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
20 – 24.9	19	DFO only	1	0.82
		DFP only	4	3.28
		DFX only	1	0.82
		DFO + DFP	11	9.02
		DFP + DFX	2	1.64
		DFO + DFX	0	0.00
25 – 29.9	14	DFO only	4	3.28
		DFP only	0	0.00
		DFX only	1	0.82
		DFO + DFP	8	6.56
		DFP + DFX	1	0.82
		DFO + DFX	0	0.00
30 – 34.9	11	DFO only	0	0.00
		DFP only	3	2.46
		DFX only	4	3.28
		DFO + DFP	3	2.46
		DFP + DFX	0	0.00
		DFO + DFX	1	0.82
35 – 39.9	5	DFO only	0	0.00
		DFP only	2	1.64
		DFX only	0	0.00
		DFO + DFP	3	2.46
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00

40 – 44.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
45 – 49.9	3	DFO only	0	0.00
		DFP only	2	1.64
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	1	0.82
		DFO + DFX	0	0.00
50 – 54.9	3	DFO only	0	0.00
		DFP only	3	2.46
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
55 – 59.9	1	DFO only	0	0.00
		DFP only	1	0.82
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
60 – 64.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
Above 65	2	DFO only	0	0.00
		DFP only	2	1.64
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
Total			122	100.00

APPENDIX M

Table M1: Distribution of Patients in Selangor According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	52	β-Thalassaemia Major	15	1.20
		HbE/β-Thalassaemia	20	1.60
		β-Thalassaemia Intermedia	4	0.32
		HbH Disease	7	0.56
		Others	6	0.48
5 - 9.9	136	β-Thalassaemia Major	21	1.68
		HbE/β-Thalassaemia	62	4.96
		β-Thalassaemia Intermedia	10	0.80
		HbH Disease	27	2.16
		Others	16	1.28
10 -14.9	129	β-Thalassaemia Major	24	1.92
		HbE/β-Thalassaemia	72	5.76
		β-Thalassaemia Intermedia	5	0.40
		HbH Disease	25	2.00
		Others	3	0.24
15 -19.9	123	β-Thalassaemia Major	21	1.68
		HbE/β-Thalassaemia	69	5.52
		β-Thalassaemia Intermedia	8	0.64
		HbH Disease	22	1.76
		Others	3	0.24
20 - 24.9	168	β-Thalassaemia Major	54	4.32
		HbE/β-Thalassaemia	61	4.88
		β-Thalassaemia Intermedia	13	1.04
		HbH Disease	36	2.88
		Others	4	0.32
25 - 29.9	158	β-Thalassaemia Major	33	2.64
		HbE/β-Thalassaemia	72	5.76
		β-Thalassaemia Intermedia	10	0.80
		HbH Disease	39	3.12
		Others	4	0.32
30 - 34.9	142	β-Thalassaemia Major	30	2.40
		HbE/β-Thalassaemia	65	5.20
		β-Thalassaemia Intermedia	8	0.64
		HbH Disease	35	2.80
		Others	4	0.32
35 - 39.9	115	β-Thalassaemia Major	16	1.28
		HbE/β-Thalassaemia	45	3.60
		β-Thalassaemia Intermedia	7	0.56
		HbH Disease	41	3.28
		Others	6	0.48
40 - 44.9	88	β-Thalassaemia Major	5	0.40
		HbE/β-Thalassaemia	37	2.96
		β-Thalassaemia Intermedia	7	0.56
		HbH Disease	35	2.80
		Others	4	0.32

45 - 49.9	51	β-Thalassaemia Major	3	0.24
		HbE/β-Thalassaemia	21	1.68
		β-Thalassaemia Intermedia	11	0.88
		HbH Disease	15	1.20
		Others	1	0.08
50 - 54.9	25	β-Thalassaemia Major	0	0.00
		HbE/β-Thalassaemia	7	0.56
		β-Thalassaemia Intermedia	3	0.24
		HbH Disease	14	1.12
		Others	1	0.08
55 - 59.9	20	β-Thalassaemia Major	0	0.00
		HbE/β-Thalassaemia	6	0.48
		β-Thalassaemia Intermedia	1	0.08
		HbH Disease	11	0.88
		Others	2	0.16
60 - 64.9	16	β-Thalassaemia Major	0	0.00
		HbE/β-Thalassaemia	8	0.64
		β-Thalassaemia Intermedia	0	0.00
		HbH Disease	8	0.64
		Others	0	0.00
Above 65	26	β-Thalassaemia Major	0	0.00
		HbE/β-Thalassaemia	2	0.16
		β-Thalassaemia Intermedia	5	0.40
		HbH Disease	18	1.44
		Others	1	0.08
Total			1249	100.00

Table M2: Distribution of Patients in Selangor According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	16	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	14	1.73
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	2	0.25
		DFO + DFP + DFX	0	0.00
5 - 9.9	84	DFO only	10	1.24
		DFP only	11	1.36
		DFX only	55	6.80
		DFO + DFP	5	0.62
		DFP + DFX	2	0.25
		DFO + DFX	1	0.12
		DFO + DFP + DFX	0	0.00
10 - 14.9	93	DFO only	10	1.24
		DFP only	11	1.36
		DFX only	52	6.43
		DFO + DFP	17	2.10
		DFP + DFX	0	0.00
		DFO + DFX	3	0.37
		DFO + DFP + DFX	0	0.00
15 - 19.9	88	DFO only	10	1.24
		DFP only	23	2.84
		DFX only	24	2.97
		DFO + DFP	25	3.09
		DFO + DFX	3	0.37
		DFP + DFX	2	0.25
		DFO + DFP + DFX	1	0.12
20-24.9	120	DFO only	7	0.87
		DFP only	45	5.56
		DFX only	9	1.11
		DFO + DFP	55	6.80
		DFO + DFP + DFX	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	4	0.49
25 - 24.9	104	DFO only	13	1.61
		DFP only	47	5.81
		DFX only	2	0.25
		DFO + DFP	36	4.45
		DFO + DFX	4	0.49
		DFP + DFX	2	0.25
		DFO + DFP + DFX	0	0.00
30 - 34.9	96	DFO only	12	1.48
		DFP only	49	6.06
		DFX only	1	0.12
		DFO + DFP	32	3.96
		DFP + DFX	1	0.12
		DFO + DFX	1	0.12
		DFO + DFP + DFX	0	0.00

35 - 39.9	66	DFO only	6	0.74
		DFP only	40	4.94
		DFX only	2	0.25
		DFO + DFP	17	2.10
		DFP + DFX	0	0.00
		DFO + DFX	1	0.12
		DFO + DFP + DFX	0	0.00
40 - 44.5	62	DFO only	1	0.12
		DFP only	48	5.93
		DFX only	1	0.12
		DFO + DFP	10	1.24
		DFP + DFX	0	0.00
		DFO + DFX	2	0.25
		DFO + DFP + DFX	0	0.00
44 - 49.9	32	DFO only	3	0.37
		DFP only	19	2.35
		DFX only	3	0.37
		DFO + DFP	6	0.74
		DFP + DFX	0	0.00
		DFO + DFX	1	0.12
		DFO + DFP + DFX	0	0.00
50 - 54.5	14	DFO only	2	0.25
		DFP only	10	1.24
		DFX only	1	0.12
		DFO + DFP	1	0.12
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	13	DFO only	0	0.00
		DFP only	9	1.11
		DFX only	1	0.12
		DFO + DFP	3	0.37
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	9	DFO only	0	0.00
		DFP only	7	0.87
		DFX only	1	0.12
		DFO + DFP	1	0.12
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	12	DFO only	0	0.00
		DFP only	9	1.11
		DFX only	3	0.37
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			809	100.00

APPENDIX N

Table N1: Distribution of Patients in Terengganu According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	31	β-Thalassaemia Major	5	1.45
		β-Thalassaemia Intermedia	2	0.58
		HbE/β-Thalassaemia	18	5.22
		HbH Disease	6	1.74
		Others	0	0.00
5 - 9.9	50	β-Thalassaemia Major	7	2.03
		β-Thalassaemia Intermedia	3	0.87
		HbE/β-Thalassaemia	28	8.12
		HbH Disease	10	2.90
		Others	2	0.58
10 - 14.9	67	β-Thalassaemia Major	12	3.48
		β-Thalassaemia Intermedia	3	0.87
		HbE/β-Thalassaemia	33	9.57
		HbH Disease	18	5.22
		Others	1	0.29
15 - 19.9	68	β-Thalassaemia Major	11	3.19
		β-Thalassaemia Intermedia	4	1.16
		HbE/β-Thalassaemia	39	11.30
		HbH Disease	14	4.06
		Others	0	0.00
20 - 24.9	37	β-Thalassaemia Major	10	2.90
		β-Thalassaemia Intermedia	1	0.29
		HbE/β-Thalassaemia	20	5.80
		HbH Disease	4	1.16
		Others	2	0.58
25 - 29.9	29	β-Thalassaemia Major	8	2.32
		β-Thalassaemia Intermedia	3	0.87
		HbE/β-Thalassaemia	15	4.35
		HbH Disease	3	0.87
		Others	0	0.00
30 - 34.9	22	β-Thalassaemia Major	4	1.16
		β-Thalassaemia Intermedia	4	1.16
		HbE/β-Thalassaemia	12	3.48
		HbH Disease	2	0.58
		Others	0	0.00
35 - 39.9	14	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	3	0.87
		HbE/β-Thalassaemia	10	2.90
		HbH Disease	0	0.00
		Others	1	0.29
40 - 44.9	8	β-Thalassaemia Major	2	0.58
		β-Thalassaemia Intermedia	1	0.29
		HbE/β-Thalassaemia	5	1.45
		HbH Disease	0	0.00
		Others	0	0.00

45 - 49.9	6	β-Thalassaemia Major	1	0.29
		β-Thalassaemia Intermedia	3	0.87
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	2	0.58
		Others	0	0.00
50 - 54.9	8	β-Thalassaemia Major	1	0.29
		β-Thalassaemia Intermedia	1	0.29
		HbE/β-Thalassaemia	3	0.87
		HbH Disease	3	0.87
		Others	0	0.00
55 - 59.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.29
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
60 and above	4	β-Thalassaemia Major	1	0.29
		β-Thalassaemia Intermedia	1	0.29
		HbE/β-Thalassaemia	1	0.29
		HbH Disease	1	0.29
		Others	0	0.00
Total			345	100.00

Table N2: Distribution of Patients in Terengganu According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 0.49	14	DFO only	1	0.42
		DFP only	0	0.00
		DFX only	13	5.44
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	34	DFO only	2	0.84
		DFP only	7	2.93
		DFX only	25	10.46
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10 - 14.9	52	DFO only	8	3.35
		DFP only	19	7.95
		DFX only	24	10.04
		DFO + DFP	1	0.42
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15 - 19.9	50	DFO only	5	2.09
		DFP only	32	13.39
		DFX only	7	2.93
		DFO + DFP	6	2.51
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20 - 24.9	24	DFO only	5	2.09
		DFP only	12	5.02
		DFX only	1	0.42
		DFO + DFP	6	2.51
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
25 - 29.9	22	DFO only	5	2.09
		DFP only	14	5.86
		DFX only	0	0.00
		DFO + DFP	3	1.26
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 34.9	15	DFO only	1	0.42
		DFP only	10	4.18
		DFX only	0	0.00
		DFO + DFP	4	1.67
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

35 - 39.9	11	DFO only	1	0.42
		DFP only	6	2.51
		DFX only	2	0.84
		DFO + DFP	2	0.84
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	7	DFO only	1	0.42
		DFP only	4	1.67
		DFX only	1	0.42
		DFO + DFP	1	0.42
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	2	DFO only	0	0.00
		DFP only	2	0.84
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	5	DFO only	1	0.42
		DFP only	3	1.26
		DFX only	0	0.00
		DFO + DFP	1	0.42
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	1	DFO only	0	0.00
		DFP only	1	0.42
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	2	DFO only	0	0.00
		DFP only	1	0.42
		DFX only	1	0.42
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			239	100.00

APPENDIX O

Table O1: Distribution of Patients in Kuala Lumpur According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 - 4.9	38	β-Thalassaemia Major	15	2.96
		β-Thalassaemia Intermedia	2	0.40
		HbE/β-Thalassaemia	13	2.57
		HbH Disease	5	0.99
		Others	3	0.59
5 - 9.9	96	β-Thalassaemia Major	17	3.36
		β-Thalassaemia Intermedia	4	0.79
		HbE/β-Thalassaemia	44	8.70
		HbH Disease	21	4.15
		Others	10	1.98
10 - 14.9	105	β-Thalassaemia Major	28	5.53
		β-Thalassaemia Intermedia	1	0.20
		HbE/β-Thalassaemia	48	9.49
		HbH Disease	19	3.75
		Others	9	1.78
15 - 19.9	95	β-Thalassaemia Major	24	4.74
		β-Thalassaemia Intermedia	3	0.59
		HbE/β-Thalassaemia	49	9.68
		HbH Disease	18	3.56
		Others	1	0.20
20 - 24.9	67	β-Thalassaemia Major	19	3.75
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	29	5.73
		HbH Disease	16	3.16
		Others	3	0.59
25 - 29.9	44	β-Thalassaemia Major	13	2.57
		β-Thalassaemia Intermedia	2	0.40
		HbE/β-Thalassaemia	17	3.36
		HbH Disease	11	2.17
		Others	1	0.20
30 - 34.9	42	β-Thalassaemia Major	15	2.96
		β-Thalassaemia Intermedia	2	0.40
		HbE/β-Thalassaemia	16	3.16
		HbH Disease	8	1.58
		Others	1	0.20
35 - 39.9	15	β-Thalassaemia Major	3	0.59
		β-Thalassaemia Intermedia	1	0.20
		HbE/β-Thalassaemia	9	1.78
		HbH Disease	2	0.40
		Others	0	0.00
40 - 44.9	2	β-Thalassaemia Major	1	0.20
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	0.20
		HbH Disease	0	0.00
		Others	0	0.00

45 - 49.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
50 - 54.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	0.20
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
55 - 59.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
60 - 64.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	1	β-Thalassaemia Major	1	0.20
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			506	100.00

Table O2: Distribution of Patients in Kuala Lumpur According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	14	DFO only	1	0.34
		DFP only	0	0.00
		DFX only	13	4.44
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	61	DFO only	9	3.07
		DFP only	1	0.34
		DFX only	48	16.38
		DFO + DFP	1	0.34
		DFP + DFX	0	0.00
		DFO + DFX	2	0.68
		DFO + DFP + DFX	0	0.00
10 - 14.9	70	DFO only	7	2.39
		DFP only	5	1.71
		DFX only	50	17.06
		DFO + DFP	3	1.02
		DFP + DFX	0	0.00
		DFO + DFX	5	1.71
		DFO + DFP + DFX	0	0.00
15 - 19.9	57	DFO only	18	6.14
		DFP only	6	2.05
		DFX only	21	7.17
		DFO + DFP	9	3.07
		DFP + DFX	0	0.00
		DFO + DFX	3	1.02
		DFO + DFP + DFX	0	0.00
20 - 24.9	34	DFO only	11	3.75
		DFP only	7	2.39
		DFX only	2	0.68
		DFO + DFP	13	4.44
		DFO + DFP + DFX	1	0.34
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
25 - 29.9	23	DFO only	4	1.37
		DFP only	8	2.73
		DFX only	1	0.34
		DFO + DFP	10	3.41
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 34.9	20	DFO only	8	2.73
		DFP only	4	1.37
		DFX only	0	0.00
		DFO + DFP	6	2.05
		DFP + DFX	2	0.68
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

35 - 39.9	11	DFO only	0	0.00
		DFP only	6	2.05
		DFX only	1	0.34
		DFO + DFP	4	1.37
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	0.34
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	1	DFO only	0	0.00
		DFP only	1	0.34
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	0.34
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			293	100.00

APPENDIX P

Table P1: Distribution of Patients in Labuan According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0 – 4.9	5	β-Thalassaemia Major	1	3.70
		β-Thalassaemia Intermedia	2	7.41
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	1	3.70
		Others	1	3.70
5 – 9.9	10	β-Thalassaemia Major	6	22.22
		β-Thalassaemia Intermedia	3	11.11
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	1	3.70
10 – 14.9	7	β-Thalassaemia Major	5	18.52
		β-Thalassaemia Intermedia	2	7.41
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
15 – 19.9	3	β-Thalassaemia Major	2	7.41
		β-Thalassaemia Intermedia	1	3.70
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
20 – 24.9	1	β-Thalassaemia Major	1	3.70
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
25 – 29.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
30 – 34.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
35 – 39.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
40 – 44.9	0	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00

45 and above	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	3.70
		HbE/β-Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			27	100.00

Table P2: Distribution of Patients in Labuan According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0 - 4.9	2	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	2	11.11
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	7	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	5	27.78
		DFO + DFP	0	0.00
		DFP + DFX	1	5.56
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	5.56
10 - 14.9	4	DFO only	1	5.56
		DFP only	0	0.00
		DFX only	2	11.11
		DFO + DFP	1	5.56
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15 - 19.9	3	DFO only	2	11.11
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	1	5.56
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20 - 24.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	5.56
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
25 - 29.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 34.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

35 - 39.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 and above	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	5.56
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			18	100.00

APPENDIX Q

Table Q1: Distribution of Patients in Putrajaya According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0-4.9	5	β-Thalassaemia Major	2	5.56
		β-Thalassaemia Intermedia	1	2.78
		HbE/β-Thalassaemia	2	5.56
		HbH Disease	0	0.00
		Others	0	0.00
5-9.9	13	β-Thalassaemia Major	4	11.11
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	8	22.22
		HbH Disease	1	2.78
		Others	0	0.00
10-14.9	12	β-Thalassaemia Major	1	2.78
		β-Thalassaemia Intermedia	2	5.56
		HbE/β-Thalassaemia	8	22.22
		HbH Disease	1	2.78
		Others	0	0.00
15-19.9	3	β-Thalassaemia Major	1	2.78
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	2	5.56
		HbH Disease	0	0.00
		Others	0	0.00
20-24.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	2.78
		HbH Disease	0	0.00
		Others	0	0.00
25 and above	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	2.78
		HbE/β-Thalassaemia	1	2.78
		HbH Disease	0	0.00
		Others	0	0.00
Total			36	100.00

Table Q2: Distribution of Patients in Putrajaya According to Type of Iron Chelator Received by Age Group

Age Group (Years)	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
0-4.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	4.35
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5-9.9	9	DFO only	2	8.70
		DFP only	0	0.00
		DFX only	7	30.43
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10-14.9	9	DFO only	2	8.70
		DFP only	0	0.00
		DFX only	7	30.43
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15-19.9	2	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	2	8.70
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20-24.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	4.35
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
25 and above	1	DFO only	0	0.00
		DFP only	1	4.35
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			23	100.00

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