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MINISTRY OF HEALTH MALAYSIA

ANNUAL REPORT

OF THE MALAYSIA
THALASSAEMIA
REGISTRY

2020



MINISTRY OF HEALTH MALAYSIA

ANNUAL REPORT OF THE MALAYSIA THALASSAEMIA REGISTRY 2020



FOREWORD

FROM THE DIRECTOR GENERAL OF HEALTH

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

The Malaysian Thalassaemia Registry (MTR) launched on the 12th May 2007 is a part of the National Thalassaemia Prevention and Control Programme. Since 2007, MTR has led the reporting of thalassaemia scenarios in the country based on detailed epidemiological and clinical data collected through 13 states and three federal territories with a total of 102 participating treatment centres. MTR is the largest thalassaemia registry in the world with 8,767 thalassaemia patients to date.

These data enable our health system to identify opportunities to improve thalassaemia prevention initiatives, enhance screening programmes and optimise treatments, clinical trials and research at the national and local levels. Early detection, screening, and diagnosis have been significantly proven to improve patients' survival rates and quality of life as well as remarkably reduced the cost and complexity of thalassaemia treatment.

However, barriers to achieving a higher standard of care need to be addressed now at the individual, health system, and governmental levels 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.

A handwritten signature in black ink, appearing to read 'Dr. Radzi', with a horizontal line underneath.

DATUK DR. MUHAMMAD RADZI BIN ABU HASSAN

The Director General of Health
Ministry of Health Malaysia

FOREWORD

FROM THE DEPUTY DIRECTOR GENERAL OF HEALTH (MEDICAL)



I would like to acknowledge the exceptional teamwork by the National Thalassaemia Registry team members who have been involved in gathering, processing and producing the data for the Malaysian Thalassaemia Registry 2020. This is the third thalassaemia registry report published by the 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 on the number of people diagnosed with thalassaemia, the different types of thalassaemia, the treatment they have 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 demonstrate differences between genders, ethnicity groups and geographical areas.

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

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



DATO' DR. ASMAYANI BINTI KHALIB

The Deputy Director General of Health (Medical)
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EXECUTIVE SUMMARY

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

The year 2020 report saw 589 new patients being added into the registry making up a total of 8,767 patients. The rising number of total patients was contributed by an increased survival number, better reporting of newly diagnosed patients as we managed to capture the less severe form of thalassaemics and, improved accessibility and compliance to iron chelators. Overall, the trend of yearly new cases especially from 2015 onwards is declining in numbers and this also reflects the positive return of the government investment for the National Thalassaemia Programme. MTR has successfully mapped the geographical distribution of thalassaemia diagnosis among the Malaysian multi-ethnicity. It is observed that β -thalassaemia major is the most common diagnosis in Sabah, predominantly in the Kadazan-Dusuns, while in Peninsular Malaysia, HbE/ β -thalassaemia is the commonest diagnosis and the majority are the Malays. A high proportion of Chinese patients were diagnosed with HbH disease in comparison to other ethnics.

The improvement in the management of iron overload and improved compliance to iron chelators especially the oral preparations most likely explained for the significant decline levels of serum ferritin observed in the past few years. The mortality rates were 8.8 (2018 and 2019) and 6.0 (2020) per 1,000 thalassaemia patients. The mortality rates were 9.3 per 1,000 TDT patients and 1.9 per 1,000 NTDT patients in 2020. These findings emphasise the importance of more effective iron chelation therapy, compliance, and a dire need for end organs iron overload monitoring through MRI T2* and clinical complications particularly endocrine complications and not only to rely on serum ferritin levels.

The MTR 2020 report includes the following new data which were not detailed in the previous annual report; i) classification of patients into transfusion-dependent thalassaemia (TDT) and non-transfusion dependent thalassaemia (NTDT), ii) MRI T2* results, iii) other organs complications in particular, endocrine complication, and iv) detailed causes of death including the organisms that caused septic death. We hope that the subsequent MTR report will incorporate complete molecular diagnoses, for accurate diagnosis and better prediction on clinical phenotype. 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.

As the overall care improved tremendously following the National Thalassaemia Program, we observed better survival among the thalassaemia patients. The median survival in 2020 was approximately 54 years for the TDT patients and 70 years for the NTDT patients. The survival analysis shows a comparable cohort with other country. The marked improvement in survival of the thalassaemics was mainly driven by a reduction in deaths due to cardiac iron overload. The most likely causes for this include the accessibility to and use of MRI T2* to identify myocardial siderosis and appropriate intensification of iron chelation therapy together with other improvements in clinical care.

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

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1.0
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 results from reduced production of one or more of the sub-units of haemoglobin that comprises both α and β globin chains. The two main categories of thalassaemia are α -thalassaemia and β -thalassaemia and, they are further divided into subcategories. β -thalassaemia major produces severe anaemia that requires life-long blood transfusion for survival. The World Health Organisation has identified 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 nationwide geographical distribution of patients, socioeconomic and clinical data including treatment outcomes is still lacking. It is estimated that about 4.5% of Malaysians are carriers (George E., 1998). The molecular defects producing β -thalassaemia are heterogeneous, and each ethnic group possesses its own specific set of mutations. β -thalassaemia is more prominent among the Malays and Kadazan Dusun ethnics, while it is rarely found in Malaysian Indians. Hb Bart's hydrops fetalis has been reported mainly in 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 when they have symptomatic anaemia, poor quality of life, compromising 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 tissue including hepatic dysfunction and failure, endocrinopathies, and cardiac dysfunction.

Due to the concern over its public health burden on the country, the Malaysian Cabinet endorsed a national comprehensive thalassaemia programme on 25th August 2004, consisting of health education and population awareness drive, screening initiative, comprehensive clinical management and establishing a Malaysian Thalassaemia Registry (MTR) database. The registry was officially launched on 12th May 2007 with an 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) and university hospitals under the Ministry of Higher Education.

The MTR database 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 core data set of essential data elements was predefined by a team of experts including clinicians and the completeness and validity of data collection was ensured by expert committee appointed by MOH, Malaysia. Site visits to ensure accuracy and completeness of the data were carried out by a team of research assistants appointed by the MOH. A regular internal audit for quality control of the MTR database is performed by the company in-charge. The web-based system is accessible to enrolled users through www.mytalasemia.net.my (MyTalasemia). It is user-friendly and can be managed from different locations. The MTR database demonstrates the value of a continuously updated registry for the surveillance of health services pertaining to thalassaemia in the country. Patient registries, which include usage of large set of data, have been reported to be helpful in mapping the functionalities and providing a positive return on investment.

1.2 DEMOGRAPHIC IN MALAYSIA

Malaysia consists of 13 states and three Federal Territories (Wilayah Persekutuan (W.P.)). Eleven states and two federal territories are located in the Peninsular Malaysia. The other two states are on the island of Borneo, and the remaining one federal territory consists of islands on the off-shore of Borneo; these are collectively referred to as East Malaysia. The list of states and federal territories in Malaysia are as follows:

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

1.3 POPULATION OF MALAYSIA

Malaysia is a fast-developing country in Southeast Asia, with a population of 32.58 million in the fourth quarter of 2020. The citizens are multi-ethnic with the majority of Bumiputera (63.35%) followed by Chinese and Indian ethnicities. Non-Malaysian citizens make up 8.9% of the population (Department of Statistics Malaysia, 2020).

The Malays 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. The Malaysian Chinese are ancestral descendants from South China, while the Malaysian Indians descended from Southern India. Indigenous people known as Orang Asli are present in the East and Peninsular Malaysia.

1.4 DATA COLLECTION

The updated MTR database includes all patients diagnosed with thalassaemia up until December 2020. Updated data from the registry was retrieved at the end of December 2020 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 MTR database. 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 through periodic data cleaning. The updated data were further verified and discussed centrally during the annual National Thalassaemia meeting held from March to April 2021 attended by experts/auditors consisting of adult and paediatric haematologists, haematopathologists, the medical development division and statisticians.

1.5 PARTICIPATING TREATMENT CENTRES







2.0

**REGISTRY
REPORT**

2.0 REGISTRY REPORT

There are 137 government hospitals including three university hospitals namely 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 102 hospitals. As of 31st December 2020, 9,641 thalassaemia patients including 874 deceased patients are registered in the MTR since year 2007.

2.1 PATIENTS DEMOGRAPHIC

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 among the siblings.

Table 2.1: Total Number of Alive Thalassaemia Patients According to Population by State in 2020

State	Total Number of Population	Number of Patients (n)	Percentage (%)
Sabah	3,908,500	1907	0.05
Selangor	6,538,100	1383	0.02
Kedah	2,185,200	903	0.04
Johor	3,781,100	660	0.02
Perak	2,510,300	602	0.02
Pulau Pinang	1,773,600	523	0.03
Kelantan	1,906,700	506	0.03
W.P. Kuala Lumpur	1,773,700	503	0.03
Pahang	1,678,600	458	0.03
Terengganu	1,259,200	364	0.03
Melaka	932700	256	0.03
Sarawak	2,816,500	254	0.01
Negeri Sembilan	1,128,800	233	0.02
Perlis	254900	134	0.05
W.P. Putrajaya	110000	45	0.04
W.P. Labuan	99600	36	0.04
Total	32,657500	8767	0.03

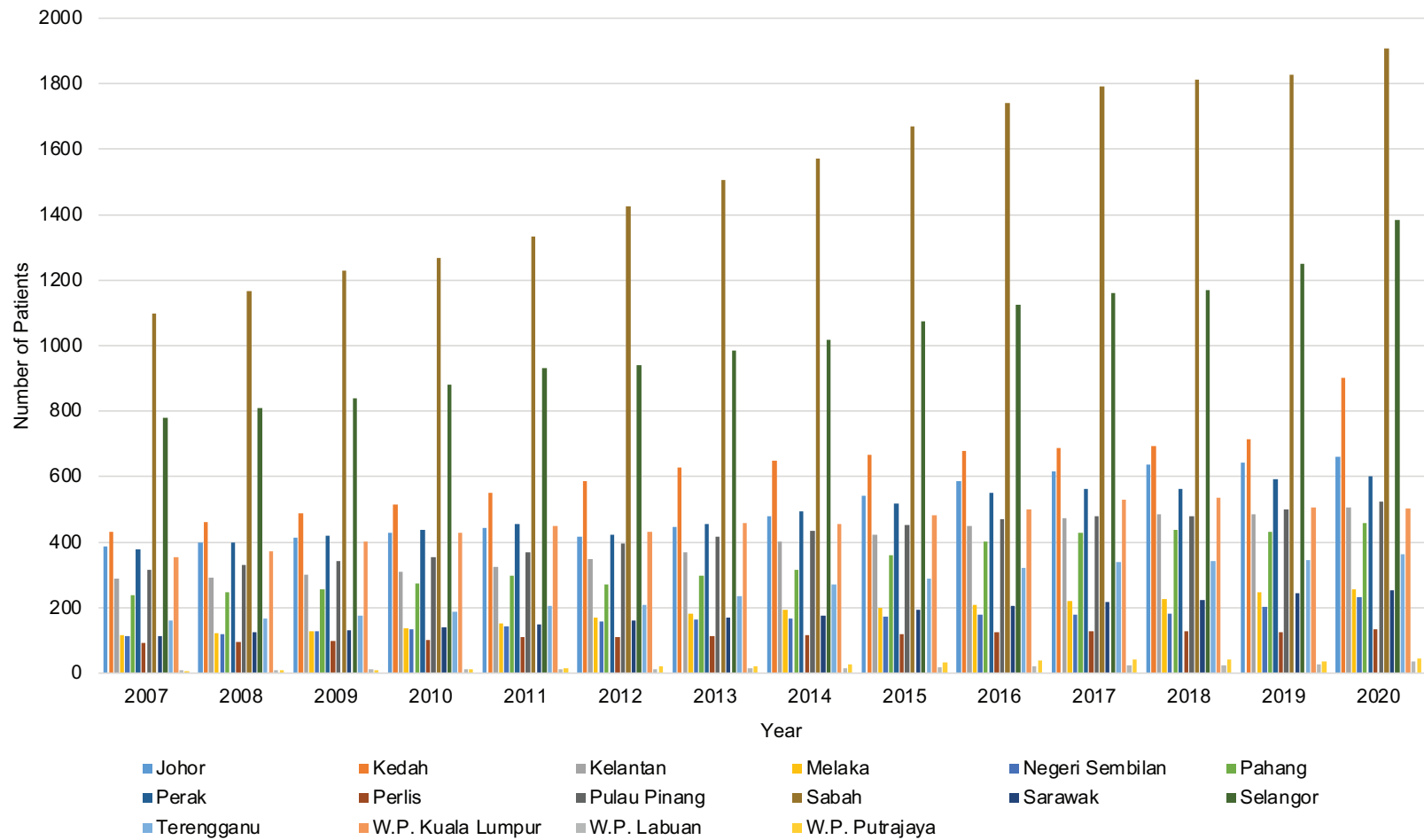


Figure 2.1: Distribution of Thalassemia Patients in Malaysia According to State by Year (2007-2020)

2.1.1 Age Groups

The mean age was 20.9 ± 13.3 years. The youngest patient in the registry is five months old with genotypic diagnosis of Hb H disease in Hospital Sungai Buloh. The oldest patient is 89 years old diagnosed with Hb H disease in Hospital Ampang. Patients in the 5–24.9 years old age group contributed to more than 50% of the total thalassaemia patients.

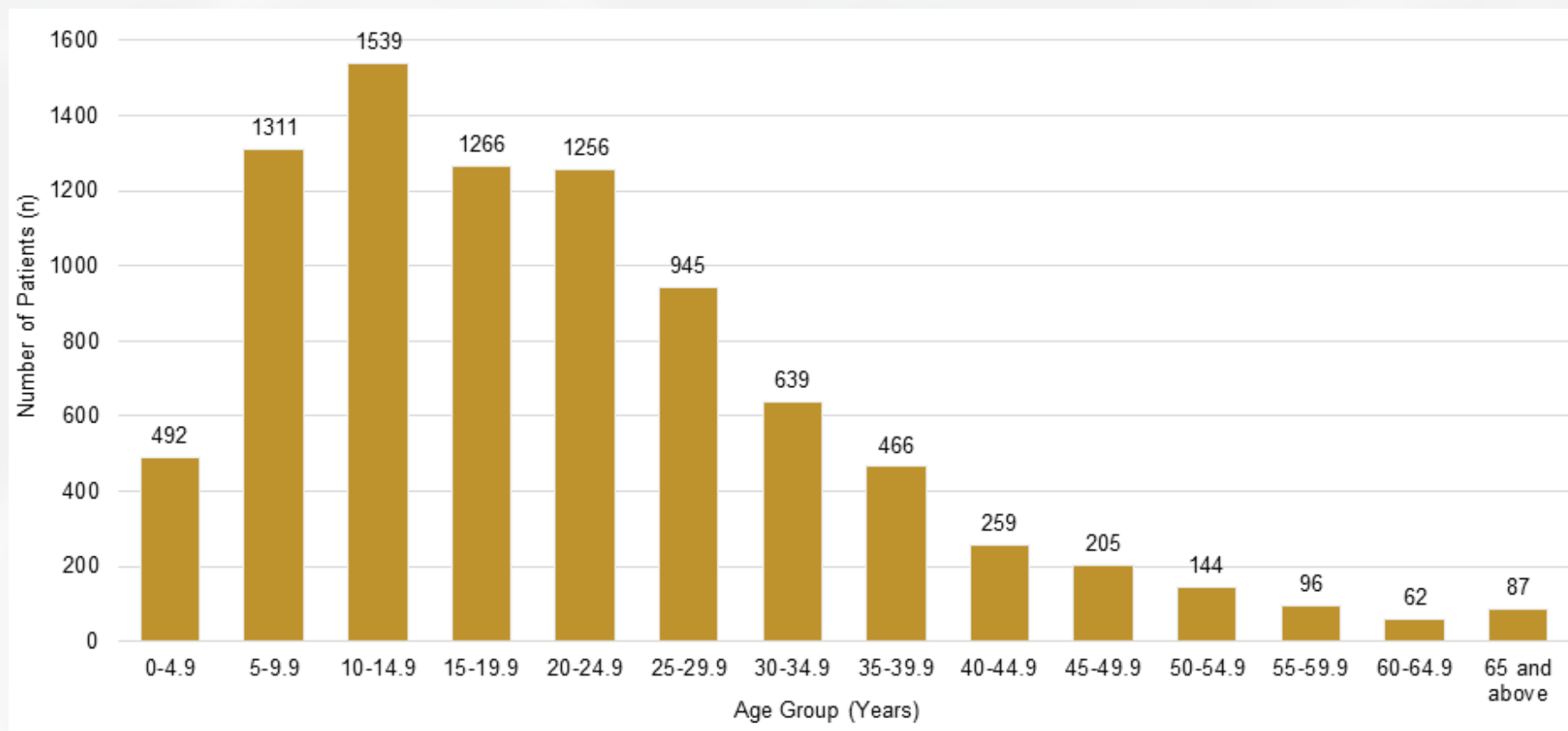


Figure 2.2: Distribution of Thalassaemia Patients in Malaysia by Age Group

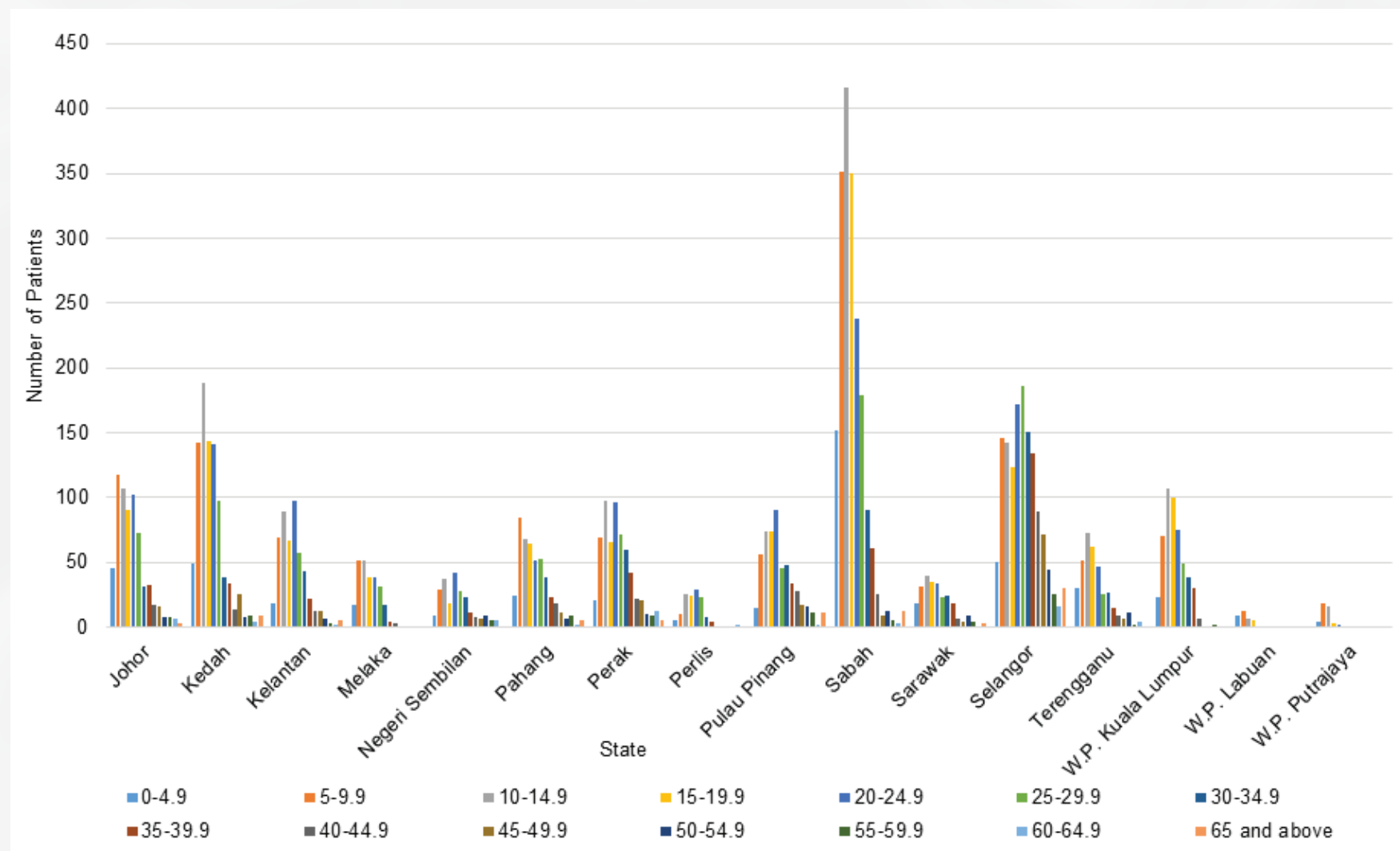


Figure 2.3: Distribution of Thalassemia Patients in Malaysia According to Age Group by State

2.1.2 Gender

The female patients were 2.3% higher than male patients based on gender distribution.

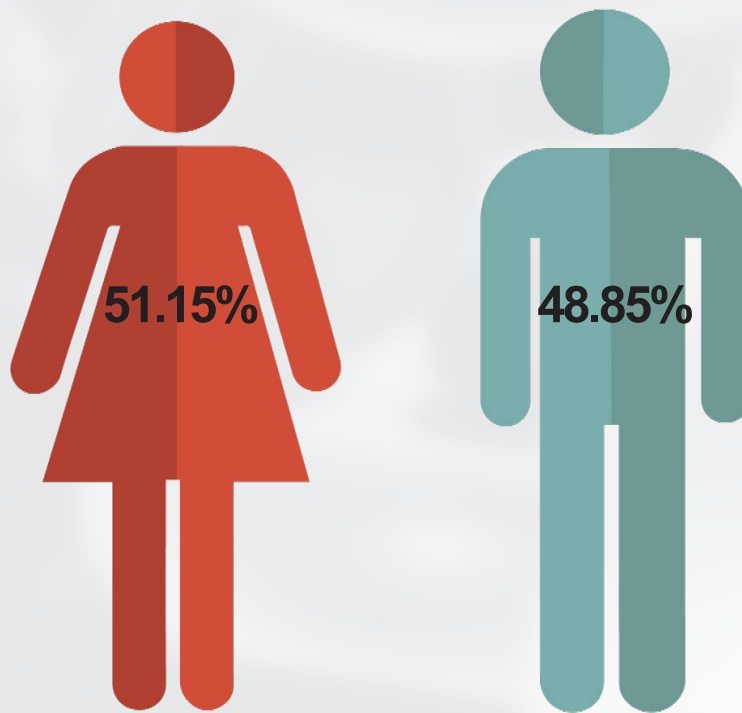


Figure 2.4: Distribution of Thalassaemia Patients in Malaysia by Gender

2.1.3 Ethnic Groups

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, the largest indigenous ethnic group in Sabah. A previous study indicated 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 the Malays formed the largest group with 5,725 patients. This is followed by the Chinese (973), Kadazan-Dusun (947). The “Others” status refer to another ethnic group which are not specified. This group forms the remaining 345 (3.94%) patients, who are either of Orang Asli, Thais, Foreigner, Iban, Pribumi Sarawak, Sino Kadazan, Kedayan, Bidayuh, Melanau, mixed or other ethnicities.

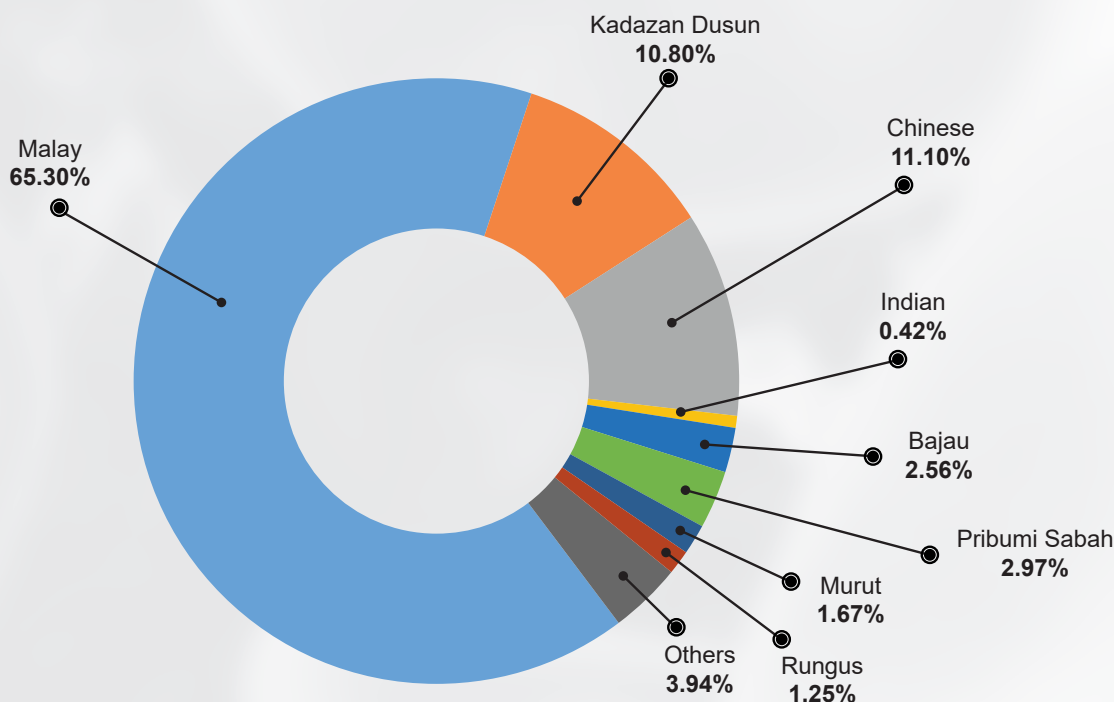


Figure 2.5: Distribution of Thalassaemia Patients in Malaysia by Ethnic Group

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 2.2: Distribution of Patients by Major Ethnic Group Based on Modified Geographical Regions

State	Total Number of Patients	Malay		Chinese		Indian		Kadazan Dusun	
		No.	%	No.	%	No.	%	No.	%
Peninsular Malaysia	6570	5495	83.64	787	11.98	36	0.55	42	0.64
Sabah + Labuan	1943	109	5.61	97	4.99	1	0.05	904	46.53
Sarawak	254	121	47.64	89	35.04	0	0.00	1	0.39
Total	8767	5725		973		37		947	

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

2.2 BIRTH SUMMARY

Overall, we observed a declining trend of new thalassaemia birth from year 2015 onwards. In 2020, there were 13 new births of thalassaemia. The new thalassaemia birth is defined as the year the patient was born. For instance, a new thalassaemia patient was diagnosed at the age of three years old in 2020 so this patient was actually the new thalassaemia birth in 2017. Given the dynamic nature of the database, the patients were only registered in the MTR after diagnosis is confirmed.

This trend could correlate with the decreasing numbers of total new cases between year 2017 and 2019. The decline is likely to reflect the success of ongoing national prevention programs, such as cascade screening and antenatal screening for pregnant mothers with anaemia. The government also embark on secondary school Form 4 thalassaemia screening which started in 2016 and this effort would contribute to further declining of new cases in the near future.

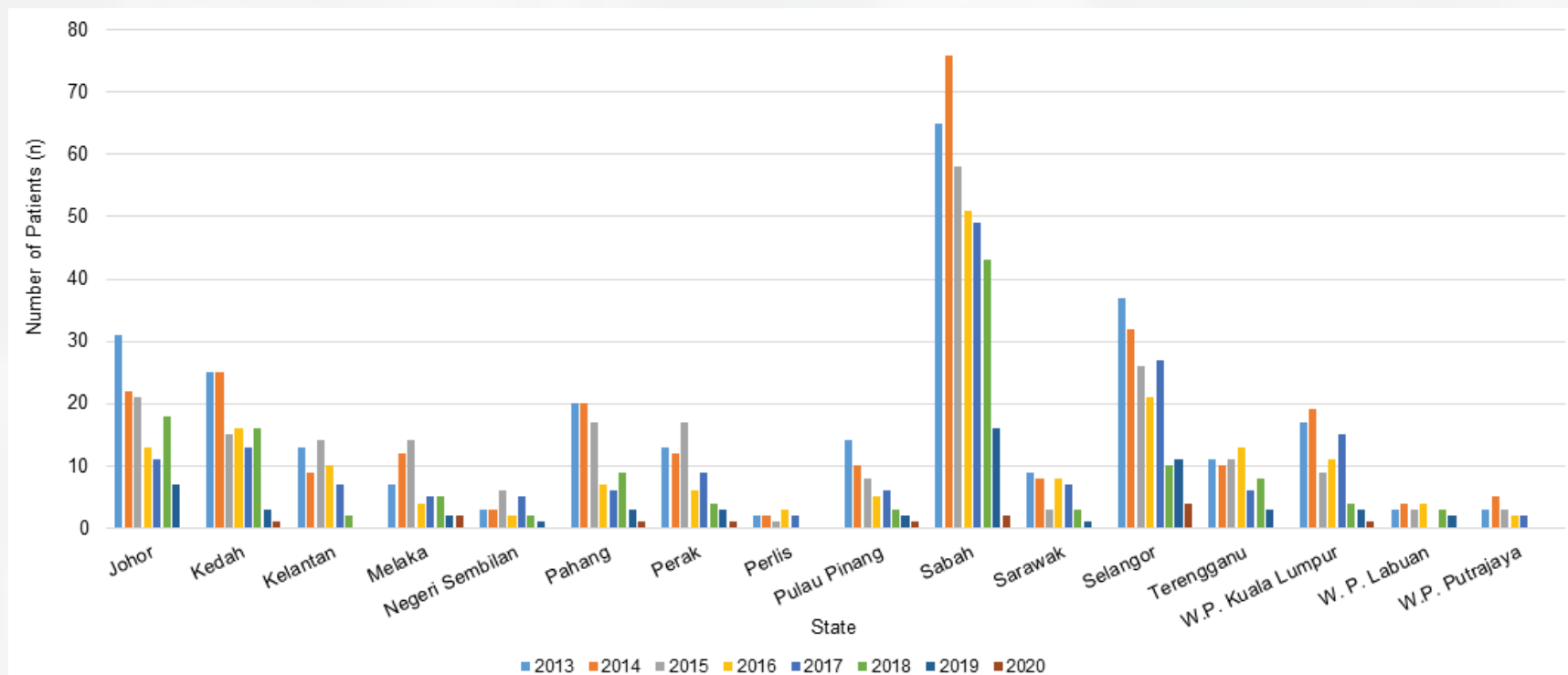


Figure 2.6: Distribution of Thalassaemia Births According to Year by State in Malaysia

2.3 DIAGNOSIS

The most common diagnosis in the registry is HbE/ β -thalassaemia followed by β -thalassaemia major, Hb H disease and β -thalassaemia intermedia. HbE/ β -thalassaemia is also the most common form of β -thalassaemia syndrome in Southeast Asia and may be presented with heterogeneous clinical condition.

The β -thalassaemia syndromes are more diverse compare to α -thalassaemia syndromes because of the diversity of mutations in the β globin genes. Patients whom diagnosed with thalassaemia intermedia may be due to mild homozygous or compound heterozygous β -globin mutation and may presented with moderately severe disease with variable transfusion requirement.

Hb H disease in this registry includes deletional type (i.e., 3 alpha gene deletions), non-deletional type (e.g., Hb H Constant Spring or Hb H Adana) and also Hb H phenotype (e.g., Hb Constant Spring coinheritance with Hb Adana).

While in the category of others, the diagnoses include Hb Lepore Hollandia, β -thalassaemia with Southeast Asia Ovalocytosis, β -thalassaemia with Hb variant, Sickle cell β -thalassaemia, Hereditary Persistence of Foetal Haemoglobin (HPFH), HbE disease, Homozygous Hb Evanton and Hb H disease with coinheritance Hb E.

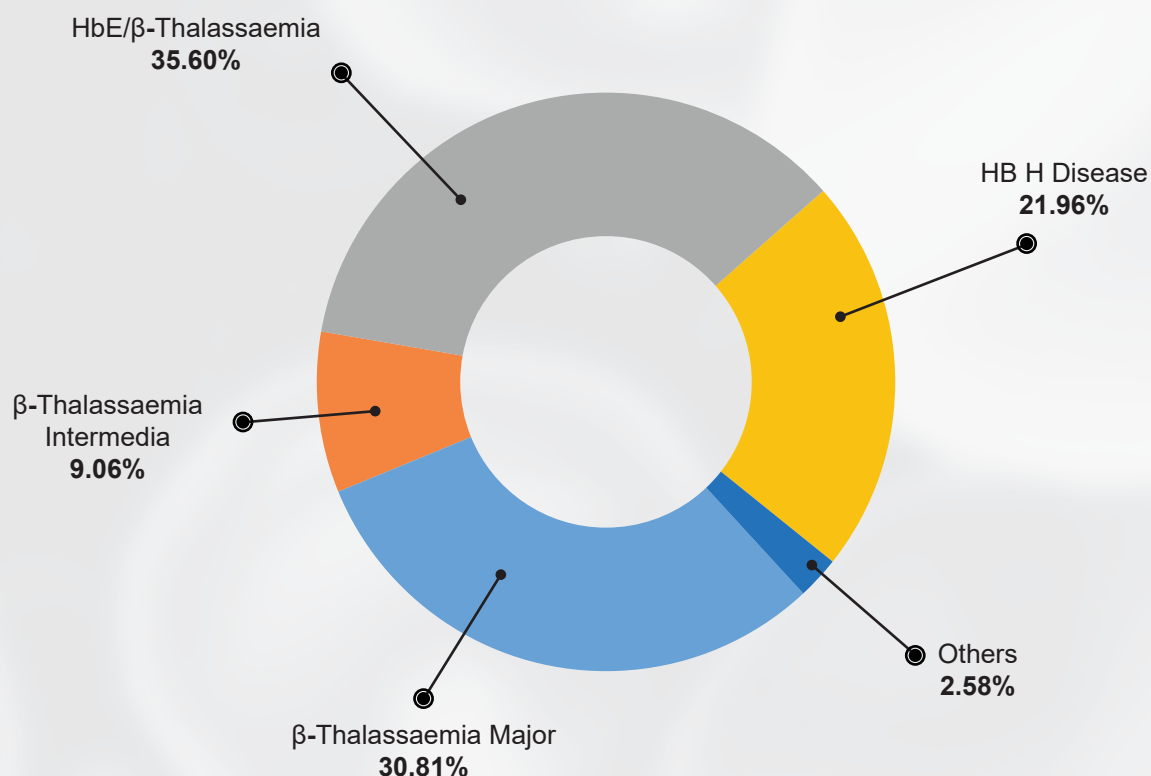


Figure 2.7: Distribution of Patients in Malaysia by Diagnosis in 2020

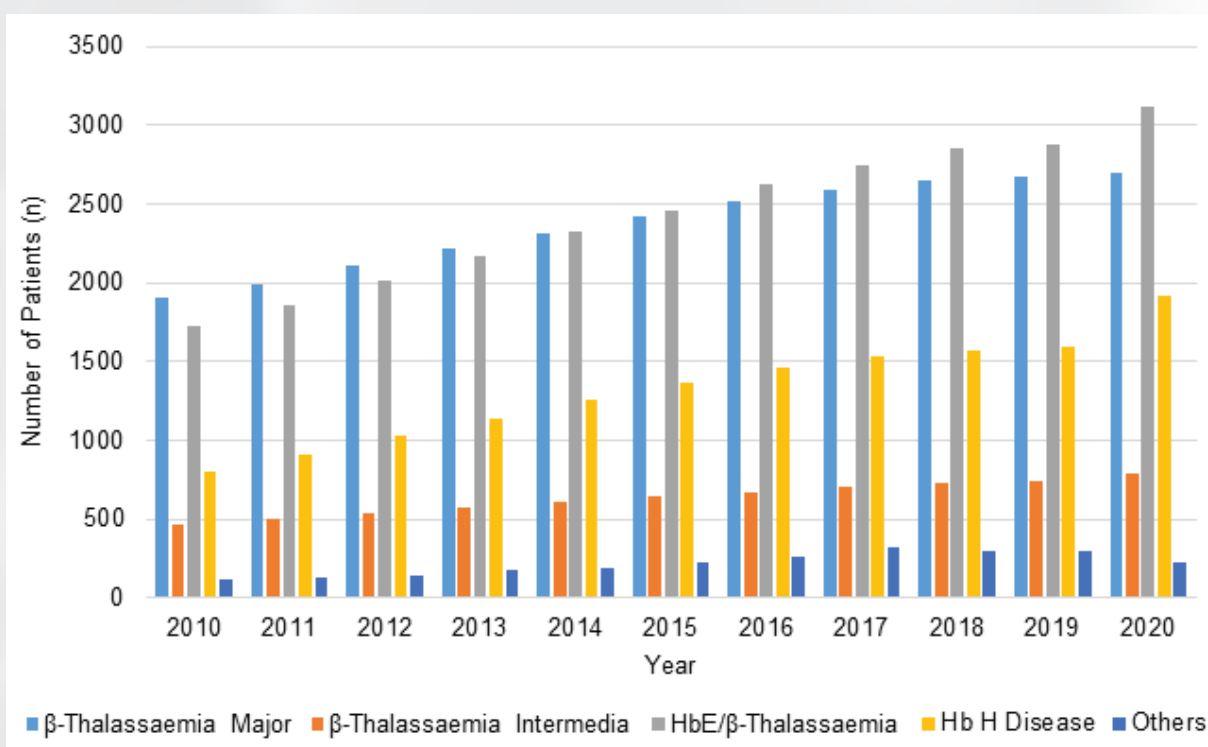


Figure 2.8: Cumulative Number of Thalassaemia Patients in Malaysia According to Diagnosis by Year

Table 2.3: Distribution of TDT and NTD T Patients in Malaysia by State

State	Total Number of Patients	Number of NTD T Patients	Percentage of NTD T Patients (%)	Number of TDT Patients	Percentage of TDT Patients (%)
Johor	660	287	43.48	373	56.52
Kedah	903	464	51.38	439	48.62
Kelantan	506	236	46.64	270	53.36
Melaka	256	119	46.48	137	53.52
Negeri Sembilan	233	130	55.79	103	44.21
Pahang	458	225	49.13	233	50.87
Perak	602	306	50.83	296	49.17
Perlis	134	51	38.06	83	61.94
Pulau Pinang	523	234	44.74	289	55.26
Sabah	1907	590	30.94	1317	69.06
Sarawak	254	138	54.33	116	45.67
Selangor	1383	694	50.18	689	49.82
Terengganu	364	115	31.59	249	68.41
W.P. Kuala Lumpur	503	249	49.50	254	50.50
W.P. Labuan	36	12	33.33	24	66.67
W.P. Putrajaya	45	16	35.56	29	64.44
Total	8767	3866	44.10	4901	55.90

The distribution of thalassaemia diagnoses in each state varies as shown in Figure 2.9

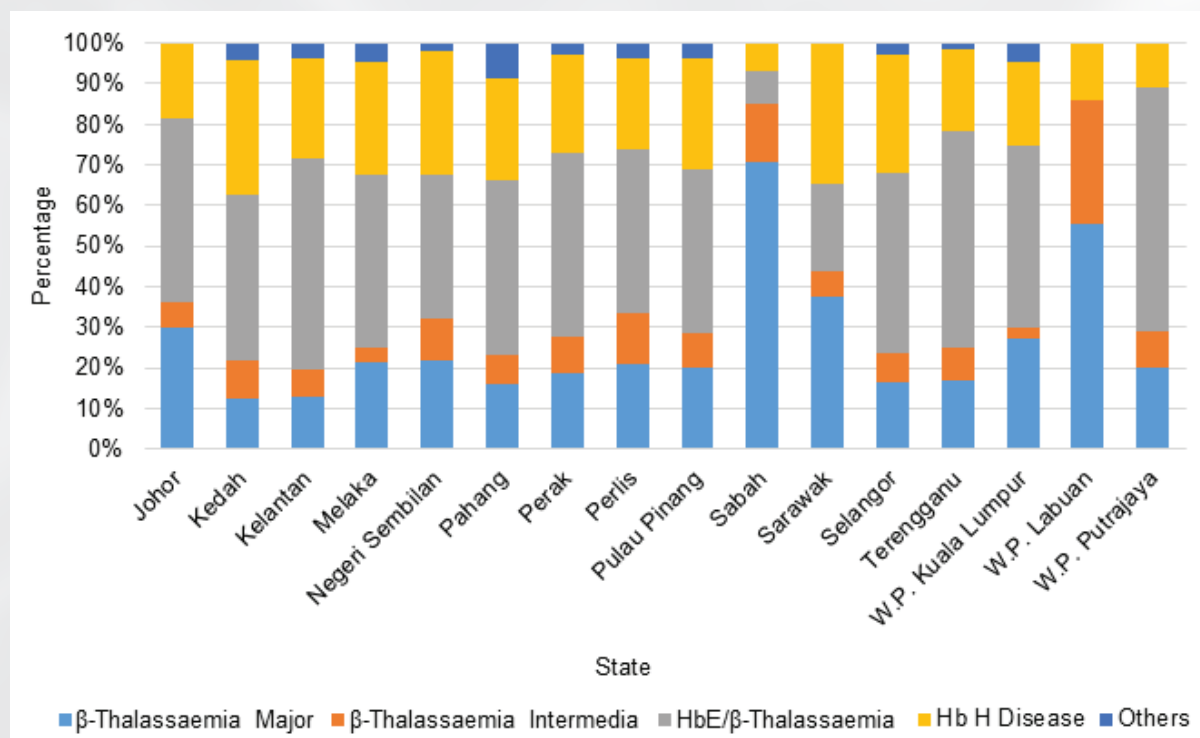


Figure 2.9: Percent Distribution of Diagnosis by State

Table 2.4 illustrates that HbE/β-thalassaemia is more common in the Malay with 2,756 patients (48.14%). Meanwhile, β-thalassaemia major is more prevalent among Chinese, Indian and Kadazan Dusun ethnicities with 392 patients (40.33%), 12 patients (31.58%), and 810 patients (83.25%), respectively.

Table 2.4: Breakdown of Diagnosis by Ethnic Group

Diagnosis	Malay		Chinese		Indian		Kadazan Dusun		Others**	
	No.	%*	No.	%*	No.	%*	No.	%*	No.	%*
β-Thalassaemia Major	947	16.54	392	40.33	12	31.58	810	83.25	540	50.99
β-Thalassaemia Intermedia	446	7.79	69	7.10	6	15.79	95	9.76	178	16.81
HbE/β-Thalassaemia	2756	48.14	134	13.79	8	21.05	33	3.39	190	17.94
Hb H Disease	1384	24.17	357	36.73	5	13.16	34	3.49	145	13.69
Others	192	3.35	20	2.06	7	18.42	1	0.10	6	0.57
Total	5725	100.00	972	100.00	38	100.00	973	100.00	1059	100.00

*Percentages are calculated to the total of each ethnic group.

**Others are mainly the ethnics of Bajau, Murut, Rungus, Pribumi Sabah and foreigners

2.4 10 CENTRES WITH HIGHEST NUMBER OF THALASSAEMIA PATIENTS

Hospital Ampang in Selangor has the highest number of thalassaemia patients among the 102 participating centres. Hospital Ampang is the national referral centre for adult haematology patients, besides being located in a high-density population, Klang Valley.

Table 2.5: 10 Centres with Highest Numbers of Thalassaemia Patients in Malaysia

Centre	Number of Patients (n)	Number of NTDT Patients	Number of TDT Patients
Hospital Ampang	757	418	339
Hospital Sultanah Bahiyah	524	281	243
Hospital Queen Elizabeth, Kota Kinabalu	378	153	225
Hospital Raja Permaisuri Bainun	360	224	136
Hospital Sultanah Aminah	322	157	165
Hospital Wanita & Kanak-Kanak Likas	318	108	210
Hospital Tengku Ampuan Rahimah	294	139	155
Hospital Pulau Pinang	286	149	137
Hospital Tengku Ampuan Afzan	270	140	130
Hospital Melaka	257	120	137

2.5 BLOOD TRANSFUSION

There are 4,901 TDT patients who received regular packed red blood cell (PRBC) transfusion, every 3-4 weekly to mitigate the chronic anaemia. The remaining 3,866 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. Many of the volume of transfusion were not captured. In future, with the collaboration with the blood bank team, it is hoped that the accurate data for transfusion volume will be available.

2.6 IRON CHELATION THERAPY

Iron removal due to transfusional iron overload is accomplished with chelating drugs, such as Desferrioxamine (DFO), Deferiprone (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 three iron chelators have established efficacy in significantly reducing iron loads from the liver and heart. Nevertheless, the indications of iron chelator 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 the 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 the faeces.

A total of 5,422 out of 8,767 (61.85%) Malaysian thalassaemia patients are on iron chelation therapy. Table 2.6 shows that 74.19% of patients are on iron chelation monotherapy while the rest are on combination therapy. DFX is usually prescribed to chelator-naive young patients and for those who could not tolerate other chelating agents. The most common combination therapy prescribed was DFO + DFP in 1,015 (18.72%) patients. The use of DFP is higher compared to last year where DFX was used in majority in our patients due to funding limitations. Based on Figure 2.11, the majority of patients on DFX are paediatric patients.

Table 2.6: Distribution of Thalassaemia Patients in Malaysia Based on Iron Chelator(s) Used

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	800	14.75
DFP only	1613	29.75
DFX only	1610	29.69
DFO + DFP	1015	18.72
DFP + DFX	178	3.28
DFO + DFX	167	3.08
DFO + DFP + DFX	39	0.72
Total	5422	100.00

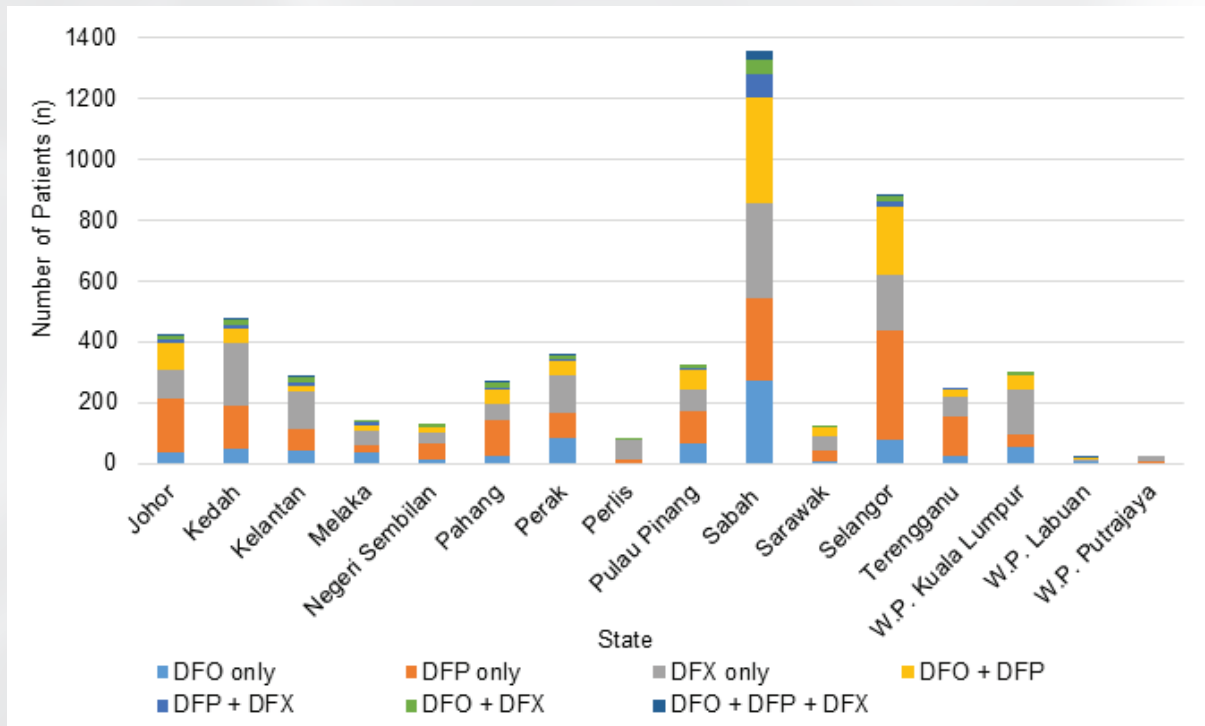


Figure 2.10: Breakdown of Patients According to Iron Chelator(s) Used by State

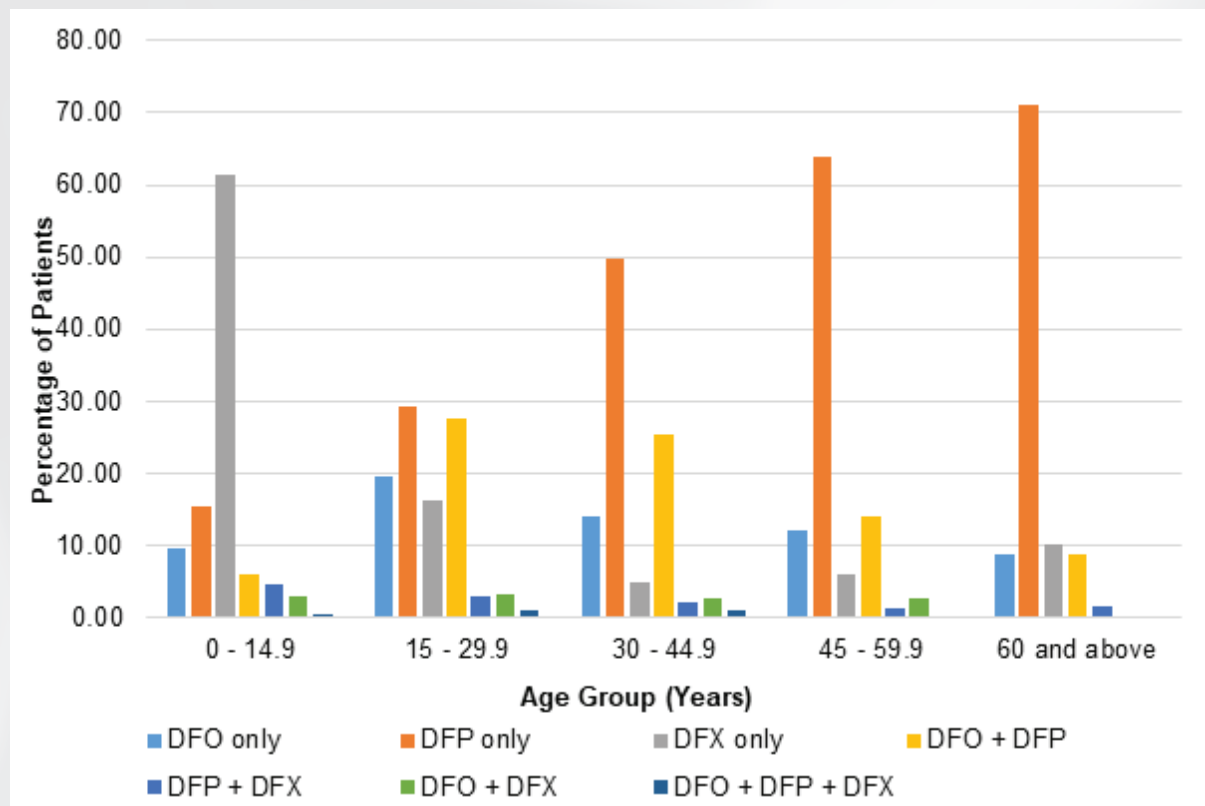


Figure 2.11: Breakdown of Patients According to Iron Chelator(s) Used by Age Group

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 manifestations of iron overload unless promptly and adequately treated with iron chelation. Therefore, the iron status should be accurately and continuously assessed to evaluate its severity, the need for chelation initiation and for monitoring the chelation therapy. Although serum ferritin levels monitoring provides a convenient method and easily accessible in all hospital but it is an acute phase reactant hence its level may change with infection and other non-iron related reasons. Therefore, it is not a reliable indicator of total body iron, liver and cardiac overload. 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 parameters 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 are now having serum ferritin levels of below 2,500 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 outcome and preservation of vital organ functions.

Table 2.7: Serum Ferritin Levels (ng/mL) in 2020

State	Total	< 1000		1000-2499		2500-4999		5000-9999		≥10,000	
		No.	%*	No.	%*	No.	%*	No.	%*	No.	%*
Johor	307	60	19.54	110	35.83	79	25.73	47	15.31	11	3.58
Kedah	323	72	22.29	140	43.34	65	20.12	31	9.60	15	4.64
Kelantan	70	11	15.71	26	37.14	21	30.00	9	12.86	3	4.29
Melaka	102	23	22.55	40	39.22	24	23.53	13	12.75	2	1.96
Negeri Sembilan	96	11	11.46	40	41.67	27	28.13	14	14.58	4	4.17
Pahang	135	37	27.41	51	37.78	30	22.22	11	8.15	6	4.44
Perak	175	28	16.00	49	28.00	69	39.43	27	15.43	2	1.14
Perlis	22	5	22.73	5	22.73	7	31.82	5	22.73	0	0.00
Pulau Pinang	148	43	29.05	45	30.41	38	25.68	17	11.49	5	3.38
Sabah	1123	299	26.63	294	26.18	259	23.06	206	18.34	65	5.79
Sarawak	116	23	19.83	56	48.28	19	16.38	15	12.93	3	2.59
Selangor	597	95	15.91	235	39.36	160	26.80	88	14.74	19	3.18
Terengganu	189	58	30.69	60	31.75	50	26.46	20	10.58	1	0.53
W.P. Kuala Lumpur	179	41	22.91	60	33.52	54	30.17	19	10.61	5	2.79
W.P. Labuan	22	3	13.64	9	40.91	8	36.36	2	9.09	0	0.00
W.P. Putrajaya	24	6	25.00	13	54.17	4	16.67	1	4.17	0	0.00
Total	3628	815	22.46	1233	33.99	914	25.19	525	14.47	141	3.89

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

There are 3,628 (41%) patients where the serum ferritin was captured in this data as compared to total patients of 8,767.

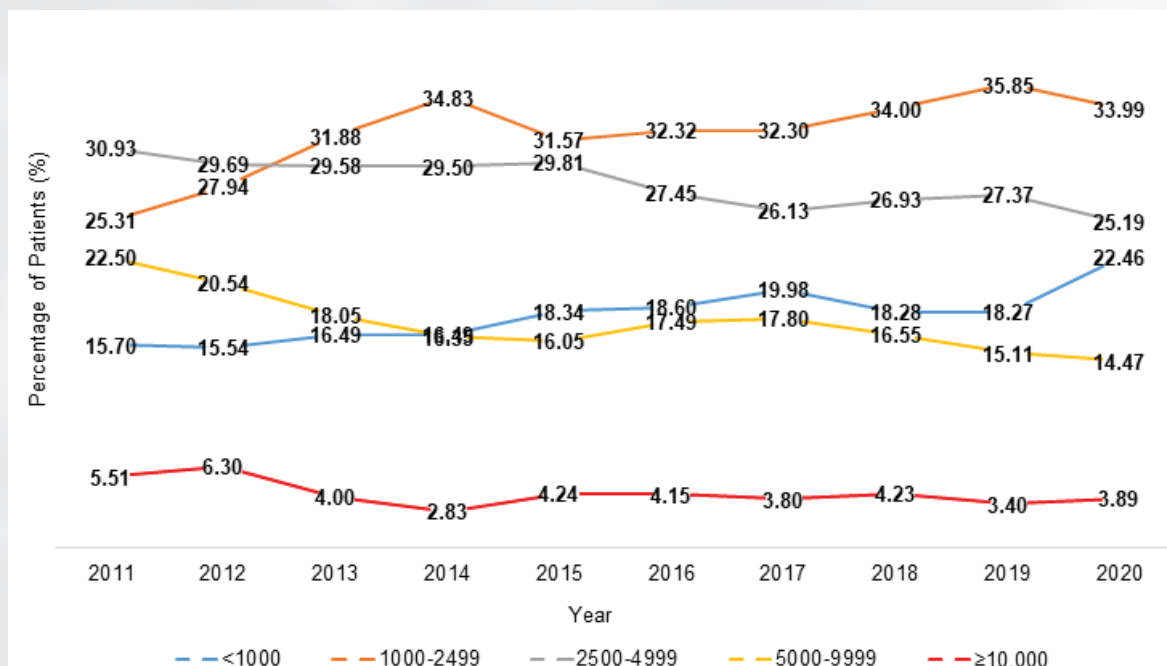


Figure 2.12: Distribution of Thalassaemia Patients in Malaysia According to Serum Ferritin Levels (ng/mL) by Year

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. In 2020, show reducing number of splenectomy procedures among the patients. Splenectomy should be delayed until the age of five years or later to avoid the risk of post-splenectomy infection (Chonat and Quinn, 2017). Out of the total 9,641 patients, 1,375 (14.26%) had undergone splenectomy. The majority of splenectomy was performed in patients diagnosed with HbE/ β -thalassaemia and β -thalassaemia major.

Table 2.8: Cumulative Number of Splenectomised Patients in Malaysia by State (2007 – 2020)

State	Total Number of Patients	Number of Patients Who Underwent Splenectomy	Percentage (%)
Johor	711	112	15.75
Kedah	945	136	14.39
Kelantan	541	117	21.63
Melaka	264	35	13.26
Negeri Sembilan	246	32	13.01

Pahang	511	113	22.11
Perak	653	92	14.09
Perlis	148	25	16.89
Pulau Pinang	541	65	12.01
Sabah	2256	235	10.42
Sarawak	279	37	13.26
Selangor	1472	242	16.44
Terengganu	394	68	17.26
W.P. Kuala Lumpur	594	63	10.61
W.P. Labuan	40	1	2.50
W.P. Putrajaya	46	2	4.35
Total	9641	1375	

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

As a comparison reported cases of splenectomy in 2019 was 47 cases, and this year, the reported cases of splenectomy was 93 cases. The increment is likely due to a better case reporting.

Table 2.9: Distribution of Splenectomised Patients in Malaysia by Diagnosis

State	β -Thalassaemia Major	β -Thalassaemia Intermedia	HbE/ β -Thalassaemia	Hb H Disease	Others
Johor	50	4	46	12	0
Kedah	49	12	57	17	1
Kelantan	29	6	72	8	2
Melaka	12	2	16	5	0
Negeri Sembilan	14	2	13	1	2
Pahang	45	11	53	4	0
Perak	29	3	53	7	0
Perlis	9	0	13	3	0
Pulau Pinang	18	11	29	5	2
Sabah	193	35	7	0	0
Sarawak	18	3	11	5	0
Selangor	73	17	127	23	2
Terengganu	22	3	40	3	0
W.P. Kuala Lumpur	21	2	37	3	0
W.P. Labuan	1	0	0	0	0
W.P. Putrajaya	0	0	2	0	0
Total	583	111	576	96	9

From clinical observations, some adult patients with NTD developed hypersplenism required multiple transfusions, therefore, splenectomy was carried out to lessen the transfusion requirement.

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	7573
Cured by Stem Cell Therapy	166
Total Living Patients	7739
Lost to Follow-up	1028
Total	8767
Cumulative Reported Deaths	874
Total	9641

Similar to 2019, Sabah has the highest cumulative number of registered thalassaemia patients in Malaysia, at 1,907 (21.75%). This is followed by Selangor (1,383), Kedah (903) and Johor at 660 patients. Meanwhile, Wilayah Persekutuan Labuan reported the lowest number of cases at 36 patients.

2.10 MORTALITY

Mortality in thalassaemia patients is 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 874 (9.07%) out of 9,641 patients cumulatively. The data demonstrated that complication due to infections was 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 Pahang, respectively. Meanwhile, Negeri Sembilan, Melaka, Putrajaya and Labuan recorded smaller number of deaths.

Out of the 874 deaths recorded, 84 had incomplete data. The leading causes of death in 790 deceased patients with verifiable data in Malaysia were infections (321 cases, 40.63%) and cardiac (311 cases, 39.37%). Figure 2.13 demonstrates the thalassaemia deaths over the past 10 years.

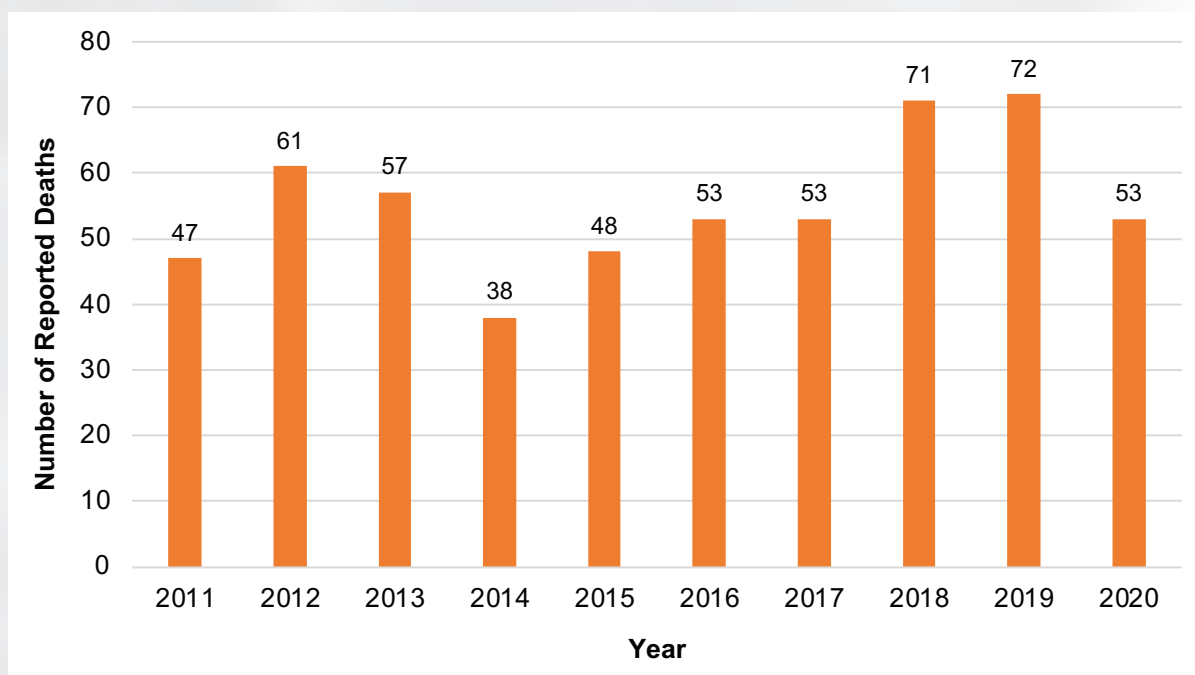


Figure 2.13: Number of Reported Deaths by Year (2011 – 2020)

Table 2.11: Cumulative Reported Deaths by State until December 2020

State	Total Number of Patients	Number of Patients (n)	Percentage over Total Thalassaemia Cases (%)
Johor	711	51	7.17
Kedah	945	42	4.44
Kelantan	541	35	6.47
Melaka	264	8	3.03
Negeri Sembilan	246	13	5.28
Pahang	511	53	10.37
Perak	653	51	7.81
Perlis	148	14	9.46
Pulau Pinang	541	18	3.33
Sabah	2256	349	15.47
Sarawak	279	25	8.96
Selangor	1472	89	6.05
Terengganu	394	30	7.61
W.P. Kuala Lumpur	594	91	15.32
W. P. Labuan	40	4	10.00
W.P. Putrajaya	46	1	2.17
Total	9641	874	9.07

The major cause of death in 2020 was infections instead of cardiac complications as in our previous report. Among the death caused by infection, six patients (25%) were splenectomised, 58% were aged between 20 to 35 years old.

Other causes of death include motor vehicle accidents (1.88%), liver disease (3.78%) and malignancy (7.55%). Another category of death in this registry is died at home/brought in dead without post mortem.

Table 2.12: Cumulative Known Cause of Death in Malaysia

Causes of Death	Cummulative Number of Patients (n)	Death in 2020	Percentage (%)
Infection	321	24	45.28
Cardiac Complications	311	18	33.96
Motor Vehicle Accidents (MVA)	23	1	1.89
Malignancy	23	4	7.55
Liver Disease	21	2	3.78
Died at Home/Brought in Dead to Hospital	17	1	1.89
Endocrine Complications	15	1	1.89
Others	15	1	1.89
Central Nervous System Event	14	1	1.89
Thalassaemia	8	0	0.00
Renal Complications	6	0	0.00
Bone Marrow Transplant Complications	6	0	0.00
Thrombosis	5	0	0.00
Surgical Complications	4	0	0.00
Acute Haemolysis	1	0	0.00
Total	790	53	100.00

Table 2.13: Distribution of Patients According to Infections Death by Age Group by Splenectomy Status in 2020

Age of Death (Years)	Total Number of Patient	Number of Patients Splenectomised (n)
0-4.9	0	0
5-9.9	0	0
10-14.9	1	0
15-19.9	3	0
20-24.9	5	1
25-29.9	6	1
30-34.9	3	3
35-39.9	0	0
40-44.9	0	0
45-49.9	2	0
50-54.9	0	0
55-59.9	2	1
60 and above	2	0
Total	24	6

2.11 COMPLICATIONS

Table 2.14 shows the cumulative transfusion transmissible infections in thalassemia patients that are reported in Malaysia. There are 102 patients with anti-HCV detected and among those only 13 patients are reported to have HCV RNA detected, 18 patients have hepatitis B surface antigen positive and three patients have anti-HIV reactive.

Table 2.14: Distribution of Patients According to Tranfusion Transmissible Infections by State

Centre	Total Number of Patients	Anti HIV	Hep B sAg	Hepatitis C: Anti HCV	Hepatitis C: HCV RNA
		No.	No.	No.	No.
Selangor	38	1	9	25	3
Terengganu	22	0	2	10	10
Perak	17	0	0	17	0
Johor	15	0	2	13	0
Pulau Pinang	13	0	0	13	0
Sarawak	12	0	2	10	0
Sabah	8	2	1	5	0
Negeri Sembilan	5	0	1	4	0
Kedah	4	0	0	4	0
Melaka	1	0	0	1	0
W.P. Kuala Lumpur	1	0	1	0	0
Kelantan	0	0	0	0	0
Pahang	0	0	0	0	0
Perlis	0	0	0	0	0
W.P. Labuan	0	0	0	0	0
W.P. Putrajaya	0	0	0	0	0
Total	136	3	18	102	13

Table 2.15: Distribution of Patients According to Endocrine Complications by State

Complications	Total Number of Patients	Short Stature	Delayed Puberty	Hypothyroid	Diabetes Mellitus	Hypogonadism
		No.	No.	No.	No.	No.
Selangor	222	97	42	49	34	0
Sarawak	98	54	27	11	6	0
Kedah	73	32	24	8	9	0
Perlis	41	14	27	0	0	0
W.P. Kuala Lumpur	32	22	5	2	2	0
Perak	31	18	8	3	2	0
Terengganu	28	0	0	12	4	12
Negeri Sembilan	22	2	2	10	8	0
Melaka	20	8	7	3	2	0
Pahang	19	9	8	1	0	1
Pulau Pinang	17	5	5	4	3	0
Johor	11	0	8	3	0	0
Kelantan	0	0	0	0	0	0
Perlis	0	0	0	0	0	0
W.P. Labuan	0	0	0	0	0	0
W.P. Putrajaya	0	0	0	0	0	0
Total	614	261	163	106	70	13

Based on Table 2.15, there are 614 patients have endocrine complication (7% of all thalassaemia patients). The most common endocrine complication is short stature (261 patients), followed by delayed puberty (163 patients), hypothyroidism (106 patients), hypogonadism (13 patients), and hypocortisol (one patient). The published data in TIF showed hypogonadism, 40%; short stature, 38% and diabetes, 9.9%.

The following Table 2.16 shows a total of 2,429 patients who had undergone MRI T2* scan to measure the iron levels in the heart. Among those, 1,931 patients (79.5%) have normal cardiac iron deposition while 250 patients (10.29%) have severe cardiac iron overload. MRI T2* usually done for patients more than 10 years old.

Table 2.16: Cardiac MRI T2* Findings by State

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Sabah	687	458	18.86	46	1.89	61	2.51	122	5.02
Selangor	604	508	20.91	23	0.95	25	1.03	48	1.98
Kedah	274	243	10.00	7	0.29	10	0.41	14	0.58
Johor	235	200	8.23	6	0.25	6	0.25	23	0.95
Negeri Sembilan	110	102	4.20	2	0.08	4	0.16	2	0.08
Terengganu	95	73	3.01	14	0.58	7	0.29	1	0.04
Perlis	73	58	2.39	8	0.33	4	0.16	3	0.12
Pulau Pinang	68	60	2.47	2	0.08	0	0.00	6	0.25
Pahang	64	57	2.35	1	0.04	1	0.04	5	0.21
Sarawak	63	51	2.10	6	0.25	0	0.00	6	0.25
Perak	62	54	2.22	1	0.04	2	0.08	5	0.21
W.P. Kuala Lumpur	52	33	1.36	5	0.21	1	0.04	13	0.54
Melaka	33	27	1.11	2	0.08	2	0.08	2	0.08
W.P. Putrajaya	4	4	0.16	0	0.00	0	0.00	0	0.00
W.P. Labuan	3	2	0.08	0	0.00	1	0.04	0	0.00
Kelantan	2	1	0.04	1	0.04	0	0.00	0	0.00
Total	2429	1931	79.50	124	5.10	124	5.10	250	10.29

Table 2.17 shows a total number of 2,388 thalassaemia patient underwent MRI scan for grading of liver iron deposition. A total of 1,073 patients (44.93%) have severe liver iron deposition compared to 240 patients (10.05%) have normal liver iron deposition.

Table 2.17: Liver MRI Findings by State

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Sabah	647	85	3.56	98	4.10	111	4.65	353	14.78
Selangor	604	41	1.72	117	4.90	159	6.66	287	12.02
Kedah	274	19	0.80	41	1.72	71	2.97	143	5.99
Johor	235	16	0.67	46	1.93	66	2.76	107	4.48
Negeri Sembilan	110	10	0.42	18	0.75	39	1.63	43	1.80
Terengganu	95	6	0.25	35	1.47	28	1.17	26	1.09
Perlis	73	6	0.25	10	0.42	16	0.67	41	1.72
Pulau Pinang	68	15	0.63	22	0.92	11	0.46	20	0.84
Pahang	64	1	0.04	11	0.46	30	1.26	22	0.92
Perak	62	10	0.42	28	1.17	20	0.84	4	0.17
Sarawak	62	5	0.21	21	0.88	28	1.17	8	0.34
W.P. Kuala Lumpur	52	22	0.92	17	0.71	11	0.46	2	0.08
Melaka	33	3	0.13	8	0.34	7	0.29	15	0.63
W.P. Putrajaya	4	1	0.04	1	0.04	1	0.04	1	0.04
W.P. Labuan	3	0	0.00	2	0.08	0	0.00	1	0.04
Kelantan	2	0	0.00	1	0.04	1	0.04	0	0.00
Total	2388	240	10.05	476	19.93	599	25.08	1073	44.93

2.12 TREATMENT ANALYSIS

2.12.1 Overall Survival of Transfusion Dependent Thalassaemia Patients by Type of Chelators

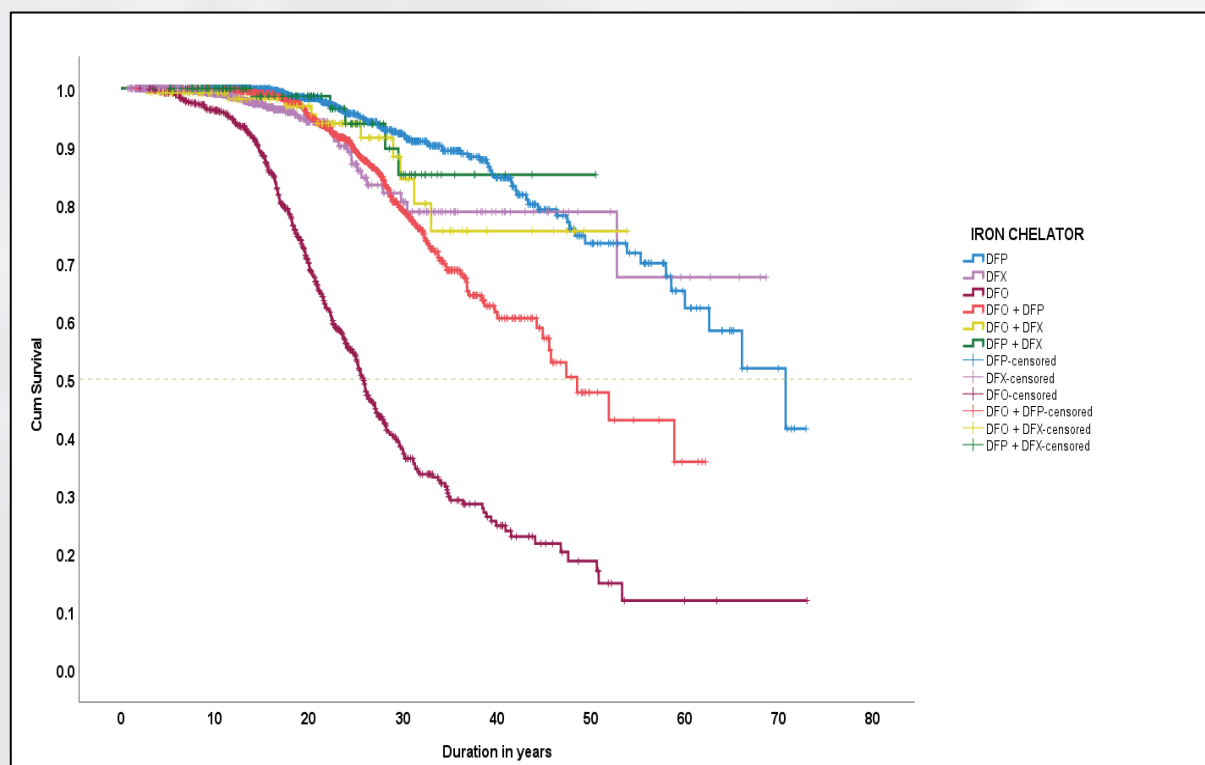


Figure 2.14: Survival of TDT Patients by Type of Chelators

The Kaplan-Meier curves show the estimated survival probability of TDT patients of six different group chelators used. Overall, survival pattern seems to be higher for patients who received combination of DFP plus DFX compared with other iron chelators. The mean estimated survival of TDT patients was calculated as 50.55 years (95% CI, 48.82 to 52.29). Patients with DFP monotherapy tended to survive longer (60.35 years; 95% CI, 57.34 to 62.97), followed by patients with DFX monotherapy (57.10 years; 95% CI, 52.47 to 61.73), while patients with DFO monotherapy have the shortest survival (31.90 years; 95% CI, 29.37 to 34.43). The log-rank test shows a significant difference in the mean survival rate between the six group of chelators [$\chi^2(5) = 615.5, p < 0.001$].

The median estimated survival time across 3 chelator groups was 70.7 years for DFP, 48.52 for DFO+DFP and 25.77 for DFO, respectively.

However, the findings of the overall survival must be interpreted cautiously, as only the ones currently using iron chelator(s) are being analysed and without adjusting other confounding factors. The type of chelator prescribed earlier was not captured by the registry.

2.12.2 Overall Survival of Transfusion Dependent Thalassaemia Patients and Non-Transfusion Dependent Thalassaemia Patients

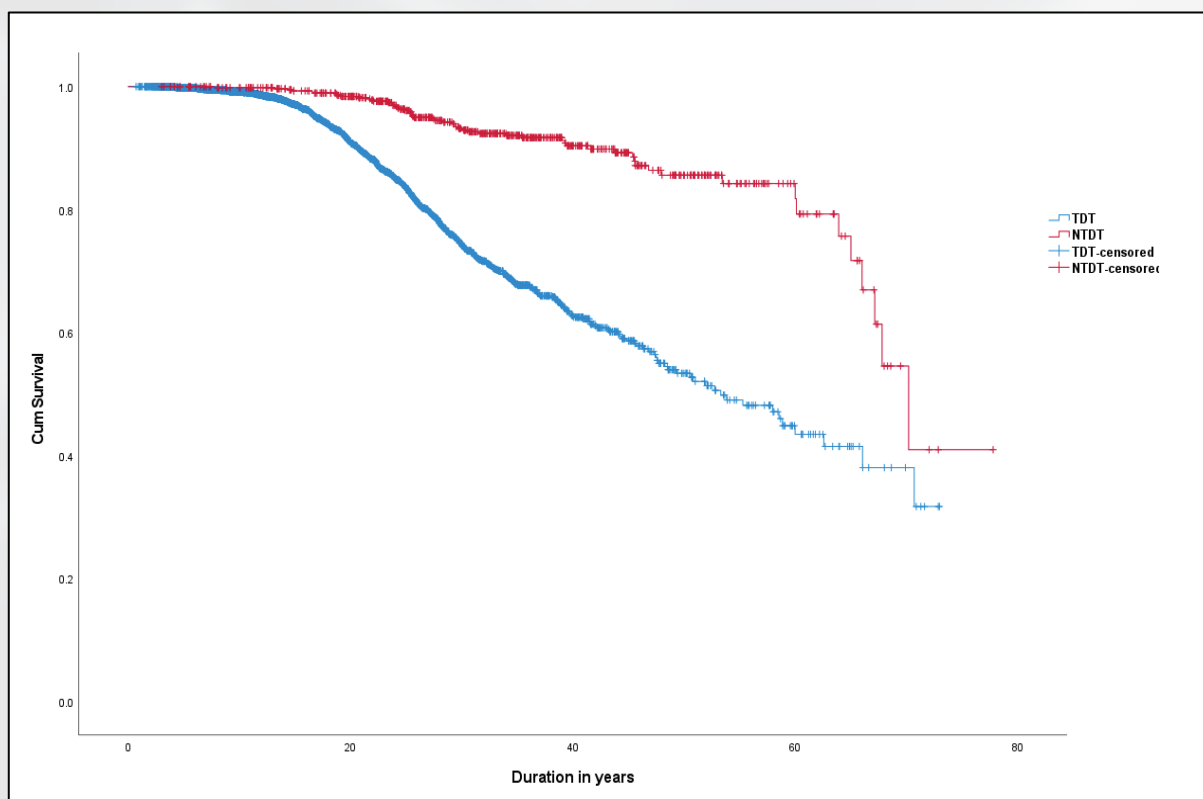


Figure 2.15: Survival of TDT and NTDT Patients

The Kaplan-Meier curves show that the NTDT patients have a higher probability of overall survival as compared to the TDT patients. At time point of 60 years, the probability of survival is approximately 0.45 (45%) for the TDT patients and 0.79 (79%) for the NTDT patients. The median survival is approximately 54 years for the TDT patients and 70 years for the NTDT patients. The log-rank test for difference in survival gives a p-value of $p = 0.001$, indicating that survival of the TDT patients and NTDT patients differ significantly.

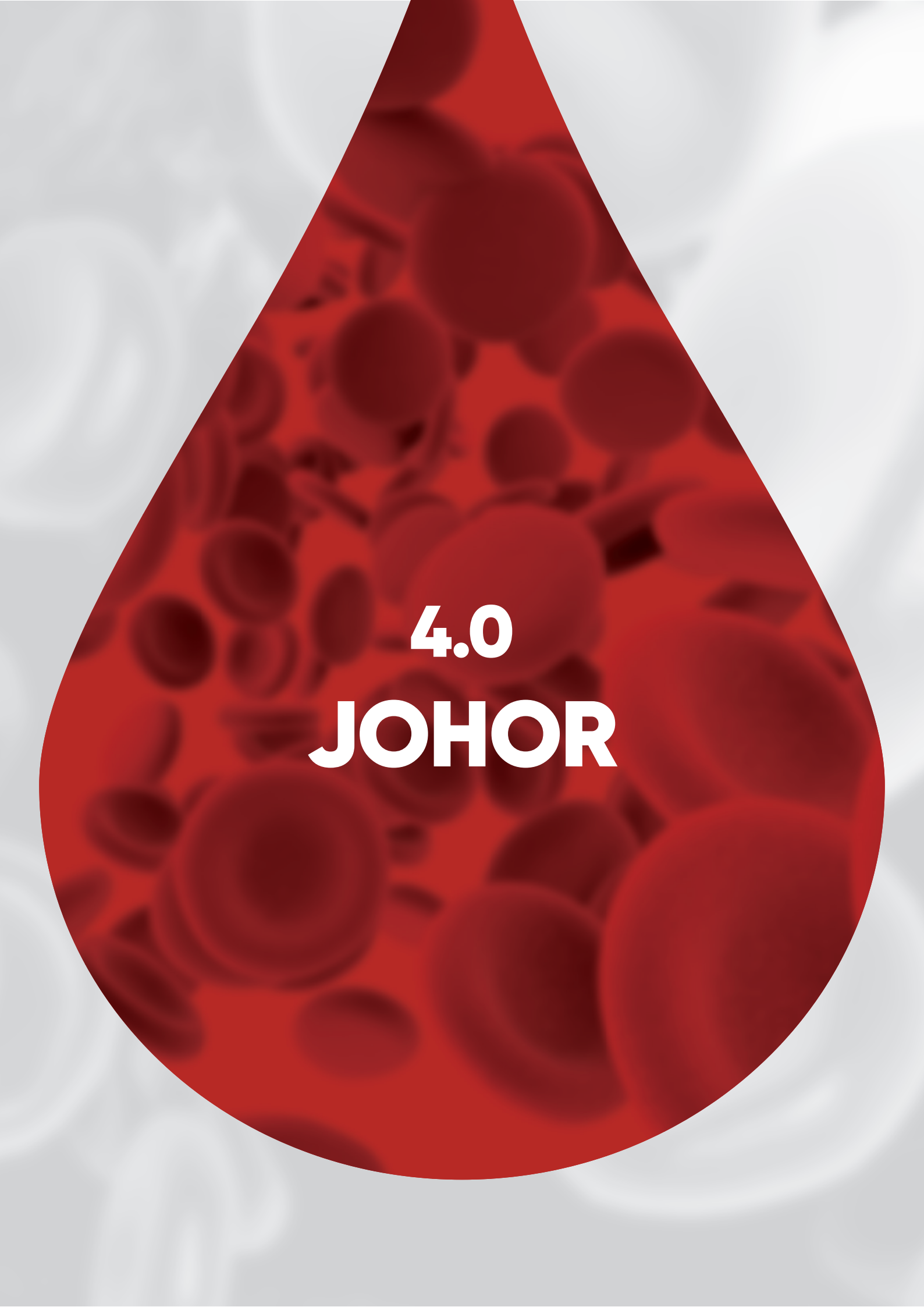


3.0
NATIONAL
CONCLUSIONS AND
OBSERVATIONS

3.0 NATIONAL CONCLUSIONS AND OBSERVATIONS

A few observations can be made from this report:

1. The total cumulative numbers of thalassaemia patients have increased by 7.2% that is 8,767 compared to 8,178 in year 2019. This is contributed by improved overall survival and better data collection and cleaning.
2. The largest number of registered patients came from the state of Sabah as shown in the previous report, followed by Selangor, Kedah, Johor and Perak.
3. Most patients in Sabah have β -thalassaemia major, whereas patients in Peninsular Malaysia are predominantly HbE/ β -thalassaemia (either TDT or NTDT). These findings can facilitate healthcare planning and resources 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 to be 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 E., 2013).
5. The number of new births and newly diagnosed cases from 2015 until present have decreased (Hishamshah I, et al. 2020). From 2014 to 2018, new thalassaemia births have declined steadily especially in Sabah. This situation may be associated with increased public awareness due to initiatives carried out by the government, in addition to the screening of secondary school children.
6. The number of patients with serum ferritin levels beyond 5,000 ng/mL is decreasing, and more patients are now having serum ferritin levels of below 2,500 ng/mL. This observation most likely indicates an improvement in iron overload management and monitoring.
7. The most common iron chelator used is oral DFP monotherapy (29.75%). As compared to the 2019 report, the most common monotherapy is DFX.
8. Newly added variables of endocrine complications in the MTR showed that we are still under-reporting.
9. We have data on MRI T2* and liver MRI to reflect the accuracy of the iron overload in the body. From MRI T2* data, there are about 20.5% who have some degree of cardiac iron overload and 90% have some degree of liver iron overload.
10. The cumulative number of patients cured by stem cell transplant is 166 and expected to increase as the transplant services are expanding in the country.
11. The accumulated number of deaths since 2007 were 874 cases. Total deaths reported in 2020 were 53 cases. The two most common accumulated causes of death were infections 45.28% (321 cases) and cardiac complications 33.96% (311 cases).
12. Overall, life expectancy seems to be higher in TDT who received combination of DFP plus DFX. The number of patients with this combination are small (3%) out of total 5422 patients on chelation.
13. The marked improvement in survival was mainly driven by a reduction in deaths due to cardiac iron overload. This is explained by the increased accessibility to and use of MRI T2* to identify myocardial siderosis and appropriate intensification of iron chelation therapy together with other improvements in clinical care.



4.0
JOHOR

4.1 INTRODUCTION

Johor is located at the southern part of Peninsular Malaysia. The total population of Johor is about 3.78 million (Department of Statistics Malaysia, 2020).

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 (HEBHK) Kluang, Hospital Sultanah Nora Ismail (HSNI) Batu Pahat, Hospital Pakar Sultanah Fatimah (HPSF) Muar, and Hospital Segamat. Hospital Permai solely treats psychiatric patients. Another five non-specialist hospitals, namely Hospital Temenggong Seri Maharaja Tun Ibrahim (HTSMTI) Kulai, Hospital Kota Tinggi, Hospital Mersing, Hospital Pontian and Hospital Tangkak.

Hospital Sultanah Aminah is a haematology centre and caters treatment for both paediatrics and adults with thalassaemia, whereas Hospital Sultan Ismail is a paediatric haematology-oncology centre treating all haematological problems, including thalassaemia for patients below 18 years old. All patients received pre-storage filtered blood from the blood bank.

4.2 PATIENT DEMOGRAPHICS

Table 4.1 shows the total number of patients alive including transplant patient are 660. Majority of patients received treatment at Hospital Sultanah Aminah (48.94%) and Hospital Sultan Ismail (14.09%), as both centres are located in Johor Bahru. Hospital Pontian and Hospital Tangkak did not have any thalassaemia patients as they are being treated at other nearest hospitals with specialists.

Table 4.1: Distribution of Patients in Johor by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Sultanah Aminah	323	48.94
Hospital Sultan Ismail	93	14.09
Hospital Pakar Sultanah Fatimah	90	13.64
Hospital Enche' Besar Hajjah Kalsom	52	7.88
Hospital Segamat	34	5.15
Hospital Mersing	34	5.15
Hospital Sultanah Nora Ismail	30	4.55
Hospital Temenggong Seri Maharaja Tun Ibrahim	3	0.45
Hospital Kota Tinggi	1	0.15
Total	660	100.00

In table 4.2, there are currently 582 living patients in Johor, 17 patients were cured by stem cell transplants and another 61 patients were lost to follow-up. There were six thalassaemia patients deaths reported in 2020.

Table 4.2: Distribution of Patients by Vital Status in Johor

Vital Status	Number of Patients (n)
Alive and On Active Treatment	582
Cured by Stem Cell Therapy	17
Total	599
Lost to Follow Up	61
Total	660
Death in 2020	6
Cumulative Reported Death	51

A total number of 17 new cases were reported in 2020. Based on Figure 4.1, there is no new thalassaemia birth in 2020.

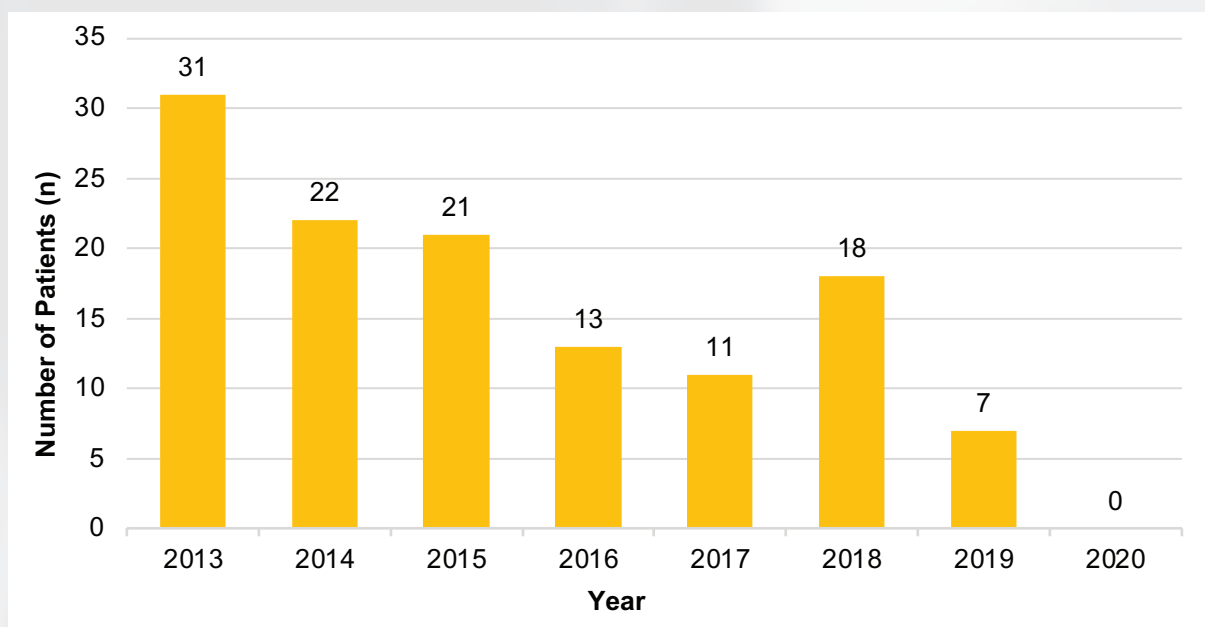


Figure 4.1: Distribution of Thalassaemia Births in Johor by Year

4.2.1 Age Groups

Majority of thalassaemia patients are aged 5 to 25 years (Figure 4.2). In all age groups, most patients were diagnosed with HbE/ β -thalassaemia (Table 4.3). The youngest patient in Johor is a one year-old boy diagnosed with β -thalassaemia major from Hospital Mersing. While, the oldest is a 71 years old male with HbH disease under follow up in Hospital Sultanah Aminah.

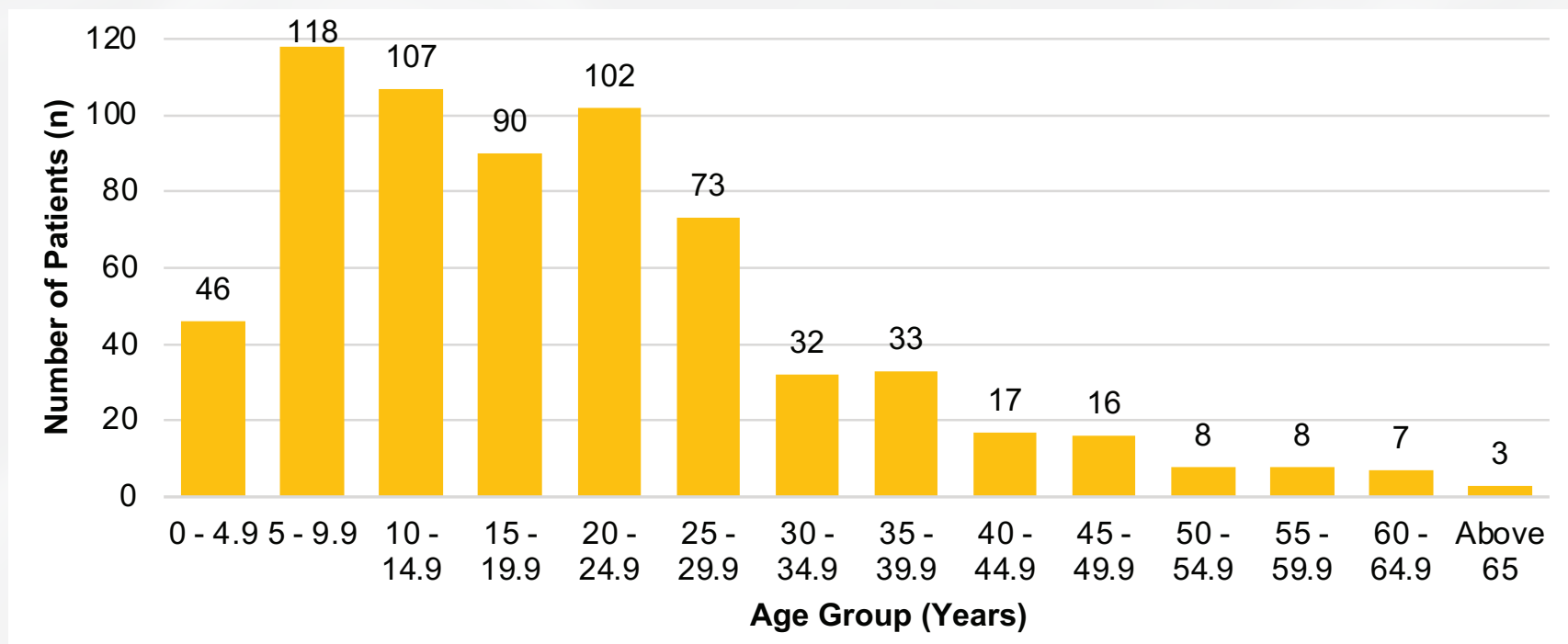


Figure 4.2: Distribution of Patients in Johor by Age Group

Table 4.3: Distribution of Patients in Johor According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0–14.9	271	β-Thalassaemia Major	75	27.68
		β-Thalassaemia Intermedia	9	3.32
		HbE/β-Thalassaemia	128	47.23
		Hb H Disease	59	21.77
		Others	0	0.00
15–29.9	265	β-Thalassaemia Major	90	33.96
		β-Thalassaemia Intermedia	12	4.53
		HbE/β-Thalassaemia	125	47.17
		Hb H Disease	38	14.34
		Others	0	0.00
30–44.9	82	β-Thalassaemia Major	24	29.27
		β-Thalassaemia Intermedia	11	13.41
		HbE/β-Thalassaemia	34	41.46
		Hb H Disease	13	15.85
		Others	0	0.00
45–59.9	32	β-Thalassaemia Major	6	18.75
		β-Thalassaemia Intermedia	7	21.88
		HbE/β-Thalassaemia	13	40.63
		Hb H Disease	6	18.75
		Others	0	0.00
Above 60	10	β-Thalassaemia Major	2	20.00
		β-Thalassaemia Intermedia	2	20.00
		HbE/β-Thalassaemia	0	0.00
		Hb H Disease	6	60.00
		Others	0	0.00
		Total	660	

4.2.2 Gender

As at 2020, there are 317 male patients (48.03%) and 343 female patients (51.97%) reported in Johor. The distribution of male and female patients in each hospital is as shown in Table 4.4.

Table 4.4: Distribution of Patients in Johor According to Gender by Centre

Centre	Total Number of Patients	Male		Female	
		No.	%	No.	%
Hospital Sultanah Aminah	323	147	22.27	176	26.67
Hospital Sultan Ismail	93	47	7.12	46	6.97
Hospital Pakar Sultanah Fatimah	90	47	7.12	43	6.52
Hospital Enche' Besar Hajjah Kalsom	52	25	3.79	27	4.09
Hospital Segamat	34	15	2.27	19	2.88
Hospital Mersing	34	15	2.27	19	2.88
Hospital Sultanah Nora Ismail	30	18	2.73	12	1.82
Hospital Temenggung Seri Maharaja Tun Ibrahim	3	3	0.45	0	0.00
Hospital Kota Tinggi	1	0	0.00	1	0.15
Total	660	317	48.03	343	51.97

4.2.3 Ethnic Group

The distribution of patients in Johor according to the major ethnic groups are Malay (501 patients, 75.91%), followed by Chinese (116 patients, 17.58%), Kadazan Dusun (15 patients, 2.27%) and Indian (nine patients, 1.36%). Other ethnicities (19 patients, 2.88%) are Pribumi Sabah other than Kadazan Dusun, Pribumi Sarawak, Orang Asli and foreigners (Rohingya, Singapore and Thailand).

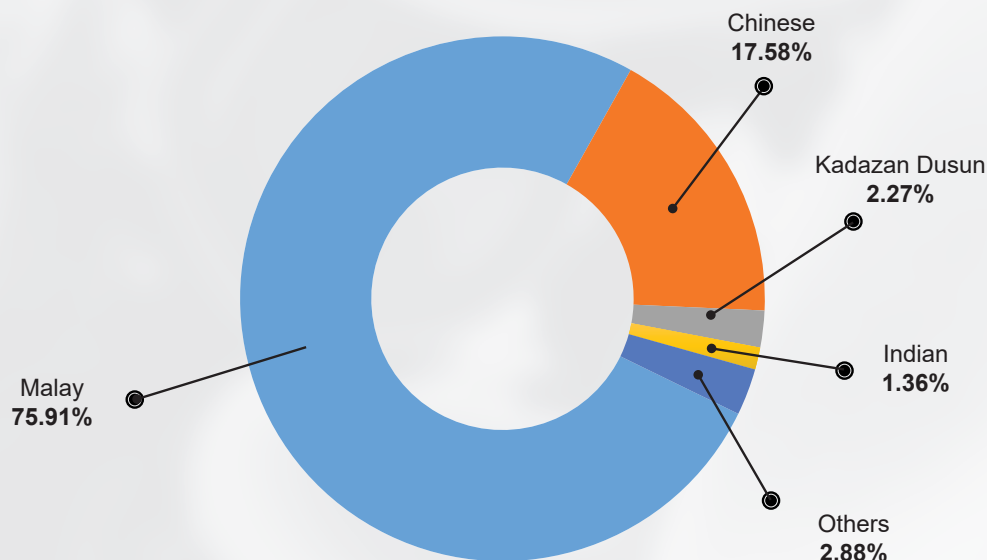


Figure 4.3: Distribution of Patients in Johor by Ethnic Group

Table 4.5: Distribution of Patients in Johor According to Ethnic Group by Centre

Centre	Total Number of Patients	Malay		Chinese		Indian		Kadazan Dusun		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Aminah	323	230	34.85	63	9.55	6	0.91	11	1.67	13	1.97
Hospital Sultan Ismail	93	63	9.55	27	4.09	0	0.00	2	0.30	1	0.15
Hospital Pakar Sultanah Fatimah	90	78	11.82	8	1.21	1	0.15	0	0.00	3	0.45
Hospital Enche' Besar Hajjah Kalsom	52	38	5.76	12	1.82	0	0.00	2	0.30	0	0.00
Hospital Segamat	34	28	4.24	3	0.45	2	0.30	0	0.00	1	0.15
Hospital Mersing	34	32	4.85	1	0.15	0	0.00	0	0.00	1	0.15
Hospital Sultanah Nora Ismail	30	28	4.24	2	0.30	0	0.00	0	0.00	0	0.00
Hospital Temenggung Seri Maharaja Tun Ibrahim	3	3	0.45	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kota Tinggi	1	1	0.15	0	0.00	0	0.00	0	0.00	0	0.00
Total	660	501	75.91	116	17.58	9	1.36	15	2.27	19	2.88

4.3 DIAGNOSIS

In Johor, HbE/ β -thalassaemia has the highest number of patients with 300 patients (45.45%), followed by 197 patients of β -thalassaemia major (29.85%), 122 patients of Hb H Disease (18.48%), and 41 patients with β -thalassaemia intermedia (6.21%) as shown in the following Figure 4.4.

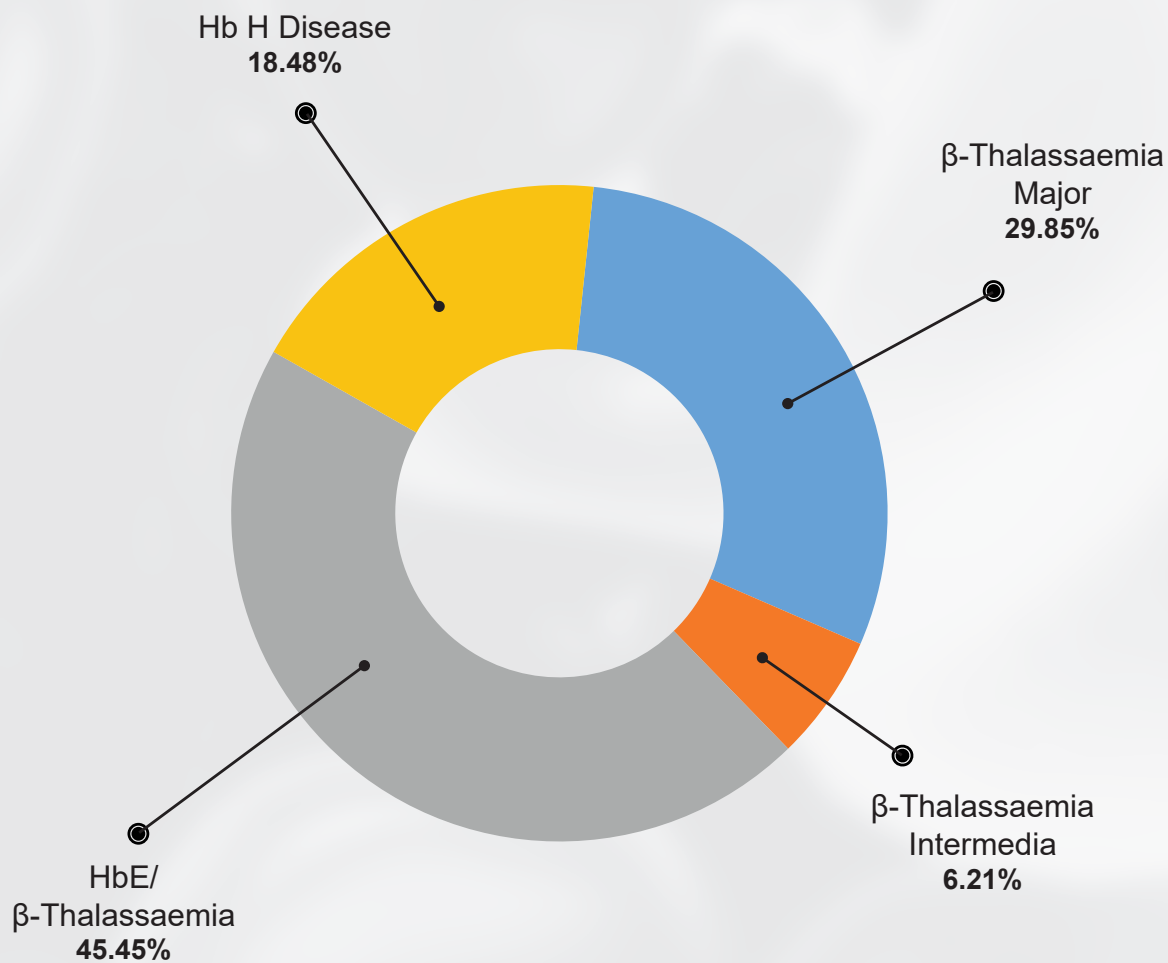


Figure 4.4: Distribution of Patients in Johor by Diagnosis

Table 4.6 shows the distribution of patients in Johor according to their diagnosis by each centre. Hospital Sultanah Aminah has the highest number of patients. The highest number of patients are diagnosed with HbE/ β -thalassaemia, followed by β -thalassaemia major except Hospital Segamat and Hospital Enche' Besar Hajjah Kalsom has slightly more β -thalassaemia major.

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

Centre	Total Number of Patients	β -Thalassaemia Major		β -Thalassaemia Intermedia		HbE/ β -Thalassaemia		Hb H Disease	
		No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Aminah	323	90	13.64	24	3.64	140	21.21	69	10.45
Hospital Sultan Ismail	93	31	4.70	2	0.30	38	5.76	22	3.33
Hospital Pakar Sultanah Fatimah	90	23	3.48	6	0.91	51	7.73	10	1.52
Hospital Enche' Besar Hajjah Kalsom	52	22	3.33	3	0.45	19	2.88	8	1.21
Hospital Segamat	34	13	1.97	4	0.61	9	1.36	8	1.21
Hospital Mersing	34	6	0.91	1	0.15	23	3.48	4	0.61
Hospital Sultanah Nora Ismail	30	8	1.21	1	0.15	20	3.03	1	0.15
Hospital Temengging Seri Maharaja Tun Ibrahim	3	3	0.45	0	0.00	0	0.00	0	0.00
Hospital Kota Tinggi	1	1	0.15	0	0.00	0	0.00	0	0.00
Total	660	197	29.85	41	6.21	300	45.45	122	18.48

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	197	Malay	125	18.94
		Chinese	50	7.58
		Indian	4	0.61
		Kadazan Dusun	12	1.82
		Others	6	0.91
β-Thalassaemia Intermedia	41	Malay	29	4.39
		Chinese	6	0.91
		Indian	2	0.30
		Kadazan Dusun	3	0.45
		Others	1	0.15
HbE/β-Thalassaemia	300	Malay	281	42.58
		Chinese	14	2.12
		Indian	0	0.00
		Kadazan Dusun	0	0.00
		Others	5	0.76
Hb H Disease	122	Malay	66	10.00
		Chinese	46	6.97
		Indian	3	0.45
		Kadazan Dusun	0	0.00
		Others	7	1.06
Total			660	100.00

Based on Table 4.7, majority of thalassaemia patients are Malay with 501 patients (75.91%) and mostly diagnosed with HbE/β-thalassaemia (300 patients, 45.45%) followed by Chinese, Kadazan Dusun and Indian.

4.4 TREATMENT

4.4.1 Iron Chelation Therapy

A total of 423 patients are on iron chelation therapy in Johor, as shown in Table 4.8. A total of 307 patients (72.58%) are on monotherapy, in which DFP with 174 patients (41.13%) contributes the highest number of patients followed by DFX (94 patients, 22.22%) and DFO (39 patients, 9.22%). A total of 115 patients (27.19%) are receiving a chelator combination of DFO and DFP as the commonest practice. Only one patient is prescribed with a combination of all three iron chelators.

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

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	39	9.22
DFP only	174	41.13
DFX only	94	22.22
DFO + DFP	91	21.51
DFP + DFX	11	2.60
DFO + DFX	13	3.07
DFO + DFP + DFX	1	0.24
Total	423	100.00

Furthermore, 340 patients (51.52%) who received iron chelation therapy are mostly those on regular blood transfusion. A majority of NTDT patients are not on iron chelation therapy. There are only 83 NTDT patients (12.58%) who are on iron chelation therapy.

Table 4.9: Distribution of Patients in Johor According to Iron Chelators by Transfusion Status

Transfusion Status	Total Number of Patients	With Iron Chelation Therapy	Percentage (%)	Without Iron Chelation Therapy	Percentage (%)
TDT	373	340	91.15	33	8.85
NTDT	287	83	28.92	204	71.08
Total	660	423	64.09	237	35.91

Table 4.10: Distribution of Patients in Johor According to Type of Iron Chelators Received by Centre

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Sultanah Aminah	186	DFO only	17	4.02
		DFP only	93	21.99
		DFX only	18	4.26
		DFO + DFP	45	10.64
		DFP + DFX	7	1.65
		DFO + DFX	5	1.18
		DFO + DFP + DFX	1	0.24
Hospital Pakar Sultanah Fatimah	67	DFO only	11	2.60
		DFP only	26	6.15
		DFX only	14	3.31
		DFO + DFP	15	3.55
		DFP + DFX	1	0.24
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Sultan Ismail	49	DFO only	1	0.24
		DFP only	13	3.07
		DFX only	31	7.33
		DFO + DFP	3	0.71
		DFP + DFX	0	0.00
		DFO + DFX	1	0.24
		DFO + DFP + DFX	0	0.00
Hospital Enche' Besar Hajjah Kalsom	40	DFO only	2	0.47
		DFP only	12	2.84
		DFX only	10	2.36
		DFO + DFP	13	3.07
		DFP + DFX	0	0.00
		DFO + DFX	3	0.71
		DFO + DFP + DFX	0	0.00
Hospital Sultanah Nora Ismail	27	DFO only	4	0.95
		DFP only	3	0.71
		DFX only	12	2.84
		DFO + DFP	7	1.65
		DFP + DFX	0	0.00
		DFO + DFX	1	0.24
		DFO + DFP + DFX	0	0.00

Hospital Mersing	27	DFO only	2	0.47
		DFP only	12	2.84
		DFX only	5	1.18
		DFO + DFP	6	1.42
		DFP + DFX	0	0.00
		DFO + DFX	2	0.47
		DFO + DFP + DFX	0	0.00
Hospital Segamat	23	DFO only	2	0.47
		DFP only	13	3.07
		DFX only	3	0.71
		DFO + DFP	1	0.24
		DFP + DFX	3	0.71
		DFO + DFX	1	0.24
		DFO + DFP + DFX	0	0.00
Hospital Temenggung Seri Maharaja Tun Ibrahim	3	DFO only	0	0.00
		DFP only	2	0.47
		DFX only	0	0.00
		DFO + DFP	1	0.24
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital KotaTinggi	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	0.24
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			423	100.00

Most of the paediatric patients at less than 10 years old are on DFX. Older patients are on Deferiprone and a few patients are on a combination therapy.

Table 4.11: 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	9	6.25
		DFP only	39	27.08
		DFX only	81	56.25
		DFO + DFP	8	5.56
		DFP + DFX	2	1.39
		DFO + DFX	5	3.47
		DFO + DFP + DFX	0	0.00
15–29.9	187	DFO only	24	12.83
		DFP only	84	44.92
		DFX only	13	6.95
		DFO + DFP	54	28.88
		DFP + DFX	7	3.74
		DFO + DFX	5	2.67
		DFO + DFP + DFX	0	0.00
30–44.9	62	DFO only	3	4.84
		DFP only	30	48.39
		DFX only	0	0.00
		DFO + DFP	23	37.10
		DFP + DFX	2	3.23
		DFO + DFX	3	4.84
		DFO + DFP + DFX	1	1.61
45–59.9	25	DFO only	3	12.00
		DFP only	17	68.00
		DFX only	0	0.00
		DFO + DFP	5	20.00
		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			423	

4.4.2 Serum Ferritin Level

There were 307 TDT patients (82.08%) had serum ferritin measured in 2020. Based on Table 4.12, 170 patients (55.37%) have serum ferritin level less than 2,500 ng/mL. However, 11 patients (3.58%) aged between 15 to 35 years old have serum ferritin above 10,000 ng/mL.

Table 4.12: 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	134	29	9.45	54	17.59	31	10.10	17	5.54	3	0.98
Hospital Sultan Ismail	47	14	4.56	19	6.19	7	2.28	5	1.63	2	0.65
Hospital Pakar Sultanah Fatimah	43	11	3.58	14	4.56	11	3.58	7	2.28	0	0.00
Hospital Enche' Besar Hajjah Kalsom	36	3	0.98	15	4.89	8	2.61	6	1.95	4	1.30
Hospital Mersing	20	2	0.65	6	1.95	5	1.63	6	1.95	1	0.33
Hospital Sultanah Nora Ismail	12	0	0.00	1	0.33	7	2.28	4	1.30	0	0.00
Hospital Segamat	11	0	0.00	1	0.33	7	2.28	2	0.65	1	0.33
Hospital Temenggung Seri Maharaja Tun Ibrahim	3	0	0.00	0	0.00	3	0.98	0	0.00	0	0.00
Hospital Kota Tinggi	1	1	0.33	0	0.00	0	0.00	0	0.00	0	0.00
Total	307	60	19.54	110	35.83	79	25.73	47	15.31	11	3.58

4.5 COMPLICATIONS AND DEATHS

4.5.1 Complications

There are 13 patients that are reported with transfusion transmissible infections of Hepatitis C and two patients were diagnosed with Hepatitis B as shown in Table 4.13. However, there is no new case of Transfusion Transmissible Infections reported in Johor for the year 2020.

Table 4.13: Distribution of Patients in Johor According to Transfusion Transmissible Infections

Complications	2007-2019	2020
Hepatitis C: Anti HCV HCV RNA	13	0
	0	0
Hepatitis B	2	0
HIV	0	0
Total	15	0

Based on Table 4.14, 11 patients had endocrine complications. It was reported that eight patients were diagnosed with delayed puberty and they are associated with higher iron deposition. In addition, three patients were diagnosed with hypothyroid.

Table 4.14: Distribution of Patients in Johor According to Endocrine Complications

Complications	Number of Patients (n)
Delayed Puberty	8
Hypothyroid	3
Total	11

4.5.2 Iron Deposition in Heart and Liver

Table 4.15 & 4.16 illustrate 235 patients had done liver and cardiac MRI T2* scan. There are 206 patients with normal, 22 patients with severe and three patients with moderate grade of cardiac iron deposition.

Table 4.15: Distribution of Patients in Johor According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Aminah	120	101	42.98	3	1.28	2	0.85	14	5.96
Hospital Pakar Sultanah Fatimah	35	28	11.91	2	0.85	1	0.43	4	1.70
Hospital Sultan Ismail	26	26	11.06	0	0.00	0	0.00	0	0.00
Hospital Enche' Besar Hajjah Kalsom	19	15	6.38	0	0.00	1	0.43	3	1.28
Hospital Sultanah Nora Ismail	17	14	5.96	1	0.43	1	0.43	1	0.43
Hospital Mersing	13	11	4.68	0	0.00	1	0.43	1	0.43
Hospital Segamat	4	4	1.70	0	0.00	0	0.00	0	0.00
Hospital Temenggung Seri Maharaja Tun Ibrahim	1	1	0.43	0	0.00	0	0.00	0	0.00
Total	235	200	85.11	6	2.55	6	2.55	23	9.79

In Table 4.16, there are 16 patients with normal, 107 patients with severe and 66 patients with moderate grade of iron deposition in their liver.

Table 4.16: Distribution of Patients in Johor According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Aminah	120	8	3.40	19	8.09	39	16.60	54	22.98
Hospital Pakar Sultanah Fatimah	35	2	0.85	6	2.55	9	3.83	18	7.66
Hospital Sultan Ismail	26	5	2.13	12	5.11	4	1.70	5	2.13
Hospital Enche' Besar Hajjah Kalsom	19	1	0.43	3	1.28	6	2.55	9	3.83
Hospital Sultanah Nora Ismail	17	0	0.00	2	0.85	6	2.55	9	3.83
Hospital Mersing	13	0	0.00	2	0.85	2	0.85	9	3.83
Hospital Segamat	4	0	0.00	2	0.85	0	0.00	2	0.85
Hospital Temenggung Seri Maharaja Tun Ibrahim	1	0	0.00	0	0.00	0	0.00	1	0.43
Total	235	16	6.81	46	19.57	66	28.09	107	45.53

4.5.3 Deaths

Based on Table 4.15, a total of 51 patients' deaths in Johor and the causes of death are as listed in Table 4.17 which includes 10 cases that are classified as unknown cause of death. There were six patients who died in 2020 with two cases for each category of infections, cardiac and malignancy.

Table 4.17: Cumulative Known Causes of Death in Johor

Causes of Death	Number of Patients (n)
Infections	24
Cardiac	10
Malignancy	5
Central Nervous System Event	2
Motor Vehicle Accident (MVA)	1
Thalassaemia	1
Others	1
Total	44

4.6 CONCLUSION

There was a total of 660 thalassaemia patients in Johor comprising 17 new thalassaemia patients. These patients are 582 patients who are alive, 17 patients were cured by stem cell transplant and 61 patients were lost to follow-up. There was no new thalassaemia birth reported in 2020.

The commonest diagnosis in Johor is HbE/ β -thalassaemia (45.45%) followed by β -thalassaemia major (29.85%), Hb H disease (18.48%) and β -thalassaemia intermedia (6.21%). There are 423 patients who are prescribed iron chelation therapy and 307 patients (72.58%) are on monotherapy, in which DFP being the most highly prescribed with 174 patients (41.13%). On the other hand, 307 (82.08%) TDT patients have their serum ferritin monitored and only 170 patients (55.37%) have serum ferritin level less than 2,500 ng/mL.

Delayed puberty is the most common endocrine complication. Out of 235 patients who had liver and cardiac MRI T2* done, 9.4% and 49% of patients have severe cardiac and liver iron loading respectively. There are 51 cumulative deaths reported in Johor and the main cause of death is due to infection. There are 6 new deaths reported in 2020 due to cardiac causes (2), infection (2) and malignancy (2). All of them were diagnosed with HbE/ β -thalassaemia except for one who was diagnosed as Hb H disease.



5.0
KEDAH

5.1 INTRODUCTION

Kedah has an estimated population of 2,185,200 in 2020 (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

Out of 903 living patients in Kedah, three patients were cured by stem cell transplant and 164 were lost to follow up. A total of 42 patients died due to several categories of cause of death. The distribution of patients in Kedah by centre is shown in Table 5.1. There are five deaths reported among thalassaemia patients in 2020.

Table 5.1: Distribution of Patients in Kedah by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Sultanah Bahiyah	525	58.14
Hospital Sultan Abdul Halim	157	17.39
Hospital Kulim	129	14.29
Hospital Sultanah Maliha	55	6.09
Hospital Kuala Nerang	16	1.77
Hospital Sik	14	1.55
Hospital Baling	7	0.78
Total	903	100.00

Table 5.2: Distribution of Patients in Kedah by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	736
Cured by Stem Cell Therapy	3
Total	739
Lost to Follow- Up	164
Total	903
Death in 2020	5
Cumulative Reported Deaths	42

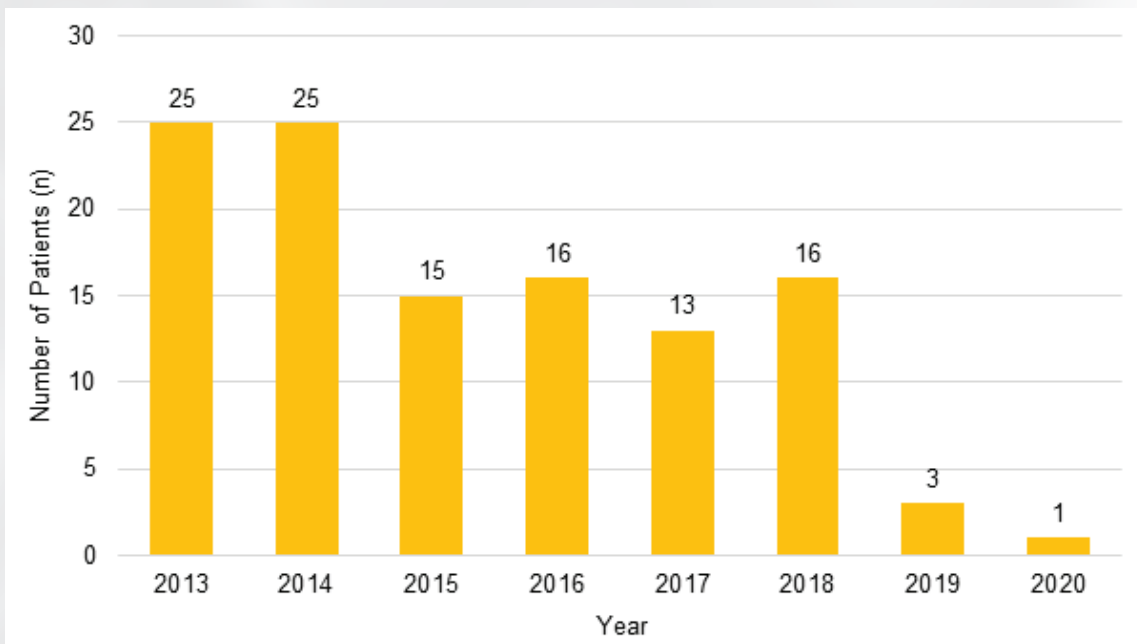


Figure 5.1: Distribution of Thalassaemia Births in Kedah by Year

5.2.1 Age Groups

A total of 522 patients (57.82%) in Kedah are between the ages of 0-19.9 years old. The youngest patient in Kedah is 9 months old (β -thalassaemia major) and the oldest is 77 years old (Delta beta ($\delta\beta$)-thalassaemia).

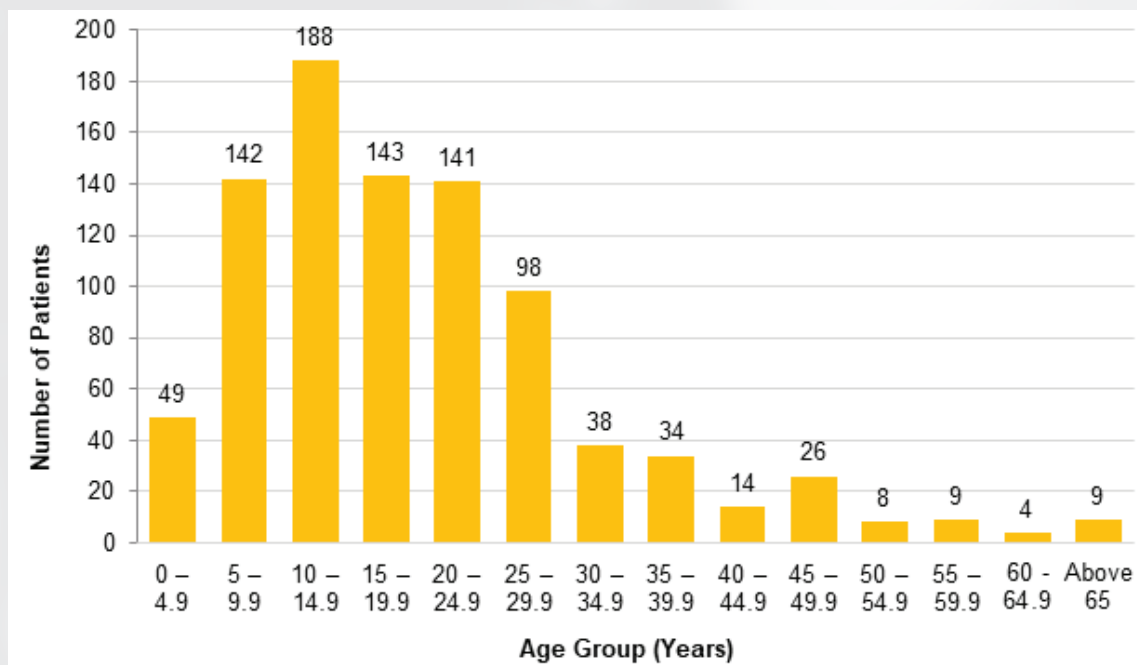


Figure 5.2: Distribution of Patients in Kedah by Age Group

Table 5.3: Distribution of Patients in Kedah According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0–14.9	379	β-Thalassaemia Major	42	11.08
		β-Thalassaemia Intermedia	42	11.08
		HbE/β-Thalassaemia	135	35.62
		Hb H Disease	142	37.47
		Others	18	4.75
15–29.9	382	β-Thalassaemia Major	57	14.92
		β-Thalassaemia Intermedia	24	6.28
		HbE/β-Thalassaemia	178	46.60
		Hb H Disease	108	28.27
		Others	15	3.93
30–44.9	86	β-Thalassaemia Major	12	13.95
		β-Thalassaemia Intermedia	10	11.63
		HbE/β-Thalassaemia	35	40.70
		Hb H Disease	27	31.40
		Others	2	2.33
45–59.9	43	β-Thalassaemia Major	2	4.65
		β-Thalassaemia Intermedia	8	18.60
		HbE/β-Thalassaemia	13	30.23
		Hb H Disease	18	41.86
		Others	2	4.65
60 and above	13	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	7.69
		HbE/β-Thalassaemia	5	38.46
		Hb H Disease	5	38.46
		Others	2	15.38
		Total	903	

5.2.2 Gender

There are 458 male patients (50.72%) and 445 female patients (49.28%) in Kedah. The distribution of male and female patients in each district hospital is shown in Table 5.4.

Table 5.4: Distribution of Patients in Kedah According to Gender by Centre

Centre	Total	Male		Female	
		No.	%	No.	%
Hospital Sultanah Bahiyah	524	248	27.46	276	30.56
Hospital Sultan Abdul Halim	157	97	10.74	60	6.64
Hospital Kulim	129	67	7.42	62	6.87
Hospital Sultanah Maliha	55	25	2.77	30	3.32
Hospital Kuala Nerang	16	7	0.78	9	1.00
Hospital Sik	14	10	1.11	4	0.44
Hospital Baling	8	4	0.44	4	0.44
Total	903	458	50.72	445	49.28

5.2.3 Ethnic Group

The distribution of patients in Kedah by ethnic group is shown in Table 5.5. Majority of the patients (849 patients, 94.02%) are Malays, 23 patients (2.55%) are Chinese, three patients (0.33%) are of Indian descent and 27 (2.99%) patients are Thais. Another patient (0.11%) is a foreigner. Figure 5.2 shows the distribution of patients in Kedah according to ethnic group by centre.

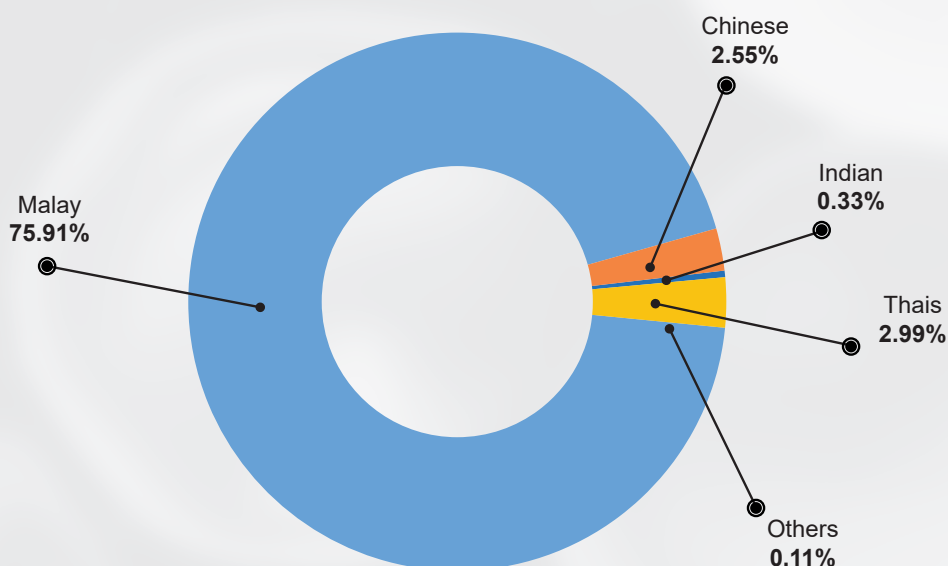


Figure 5.3: Distribution of Patients in Kedah by Ethnic Group

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

Centre	Total Number of Patients	Malay		Chinese		Indian		Thais		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Bahiyah	524	487	53.93	14	1.55	1	0.11	22	2.44	0	0.00
Hospital Sultan Abdul Halim	157	149	16.50	5	0.55	1	0.11	2	0.22	0	0.00
Hospital Kulim	129	125	13.84	2	0.22	1	0.11	0	0.00	1	0.11
Hospital Sultanah Maliha	55	53	5.87	2	0.22	0	0.00	0	0.00	0	0.00
Hospital Kuala Nerang	16	14	1.55	0	0.00	0	0.00	2	0.22	0	0.00
Hospital Sik	14	13	1.44	0	0.00	0	0.00	1	0.11	0	0.00
Hospital Baling	8	8	0.89	0	0.00	0	0.00	0	0.00	0	0.00
Total	903	849	94.02	23	2.55	3	0.33	27	2.99	1	0.11

5.3 DIAGNOSIS

HbE/ β -thalassaemia is the most common type of thalassaemia in Kedah (366 patients, 40.53%) followed by HbH disease (300 patients, 33.22%), β -thalassaemia major (113 patients, 12.51%), and β -thalassaemia intermedia (85 patients, 9.41%). Thirty-nine patients have other diagnoses (4.32%).

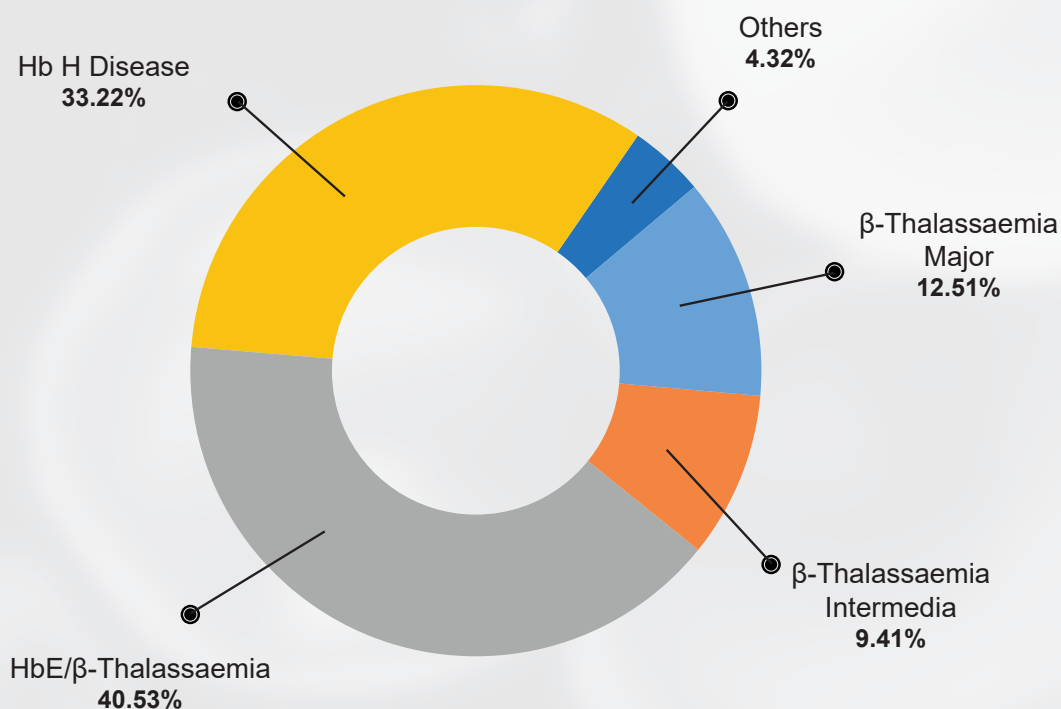


Figure 5.4: Distribution of Patients in Kedah by Diagnosis

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

Centre	Total Number of Patients	β-Thalassaemia major		β-Thalassaemia intermedia		HbE/β-Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Bahiyah	524	65	7.20	50	5.54	198	21.93	192	21.26	19	2.10
Hospital Sultan Abdul Halim	157	24	2.66	13	1.44	75	8.31	41	4.54	4	0.44
Hospital Kulim	129	11	1.22	15	1.66	55	6.09	39	4.32	9	1.00
Hospital Sultanah Maliha	55	4	0.44	5	0.55	19	2.10	20	2.21	7	0.78
Hospital Kuala Nerang	16	5	0.55	1	0.11	6	0.66	4	0.44	0	0.00
Hospital Sik	14	1	0.11	1	0.11	9	1.00	3	0.33	0	0.00
Hospital Baling	8	3	0.33	0	0.00	4	0.44	1	0.11	0	0.00
Total	903	113	12.51	85	9.41	366	40.53	300	33.22	39	4.32

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

Diagnosis	Total Number of Patients	Ethnicity	Number of Patients (n)	Percentage (%)
β- Thalassaemia Major	113	Malay	104	11.52
		Chinese	4	0.44
		Indian	1	0.11
		Thais	4	0.44
		Others	0	0.00
β- Thalassaemia Intermedia	85	Malay	81	8.97
		Chinese	4	0.44
		Indian	0	0.00
		Thais	0	0.00
		Others	0	0.00
HbE/β-Thalassaemia	366	Malay	349	38.65
		Chinese	10	1.11
		Indian	2	0.22
		Thais	5	0.55
		Others	0	0.00
Hb H Disease	300	Malay	276	30.56
		Chinese	5	0.55
		Indian	0	0.00
		Thais	18	1.99
		Others	1	0.11
Others	39	Malay	39	4.32
		Chinese	0	0.00
		Indian	0	0.00
		Thais	0	0.00
		Others	0	0.00
Total			903	100.00

5.4 TREATMENT

5.4.1 Iron Chelation Therapy

Of the 472 patients on chelation in Kedah, 43.64% are on oral DFX, 16.73% are on combination therapy, 10.59% are on subcutaneous DFO and 29.03% are on oral DFP.

Table 5.8: Distribution of Patients in Kedah by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	50	10.59
DFP only	137	29.03
DFX only	206	43.64
DFO + DFP	50	10.59
DFP + DFX	12	2.54
DFO + DFX	15	3.18
DFO+ DFP + DFX	2	0.42
Total	472	100.00

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

Centre	Total Number of Patients	Iron Chelator	Number of Patients (n)	Percentage (%)
Hospital Sultanah Bahiyah	259	DFO only	17	3.60
		DFP only	84	17.80
		DFX only	112	23.73
		DFO + DFP	25	5.30
		DFP + DFX	10	2.12
		DFO + DFX	9	1.91
		DFO+ DFP + DFX	2	0.42
Hospital Sultan Abdul Halim	87	DFO only	22	4.66
		DFP only	8	1.69
		DFX only	48	10.17
		DFO + DFP	8	1.69
		DFP + DFX	0	0.00
		DFO + DFX	1	0.21
		DFO+ DFP + DFX	0	0.00
Hospital Kulim	71	DFO only	11	2.33
		DFP only	30	6.36
		DFX only	17	3.60
		DFO + DFP	10	2.12
		DFP + DFX	1	0.21
		DFO + DFX	2	0.42
		DFO+ DFP + DFX	0	0.00
Hospital Sultanah Maliha	29	DFO only	0	0.00
		DFP only	6	1.27
		DFX only	19	4.03
		DFO + DFP	2	0.42
		DFP + DFX	0	0.00
		DFO + DFX	2	0.42
		DFO+ DFP + DFX	0	0.00
Hospital Kuala Nerang	15	DFO only	0	0.00
		DFP only	6	1.27
		DFX only	2	0.42
		DFO + DFP	5	1.06
		DFP + DFX	1	0.21
		DFO + DFX	1	0.21
		DFO+ DFP + DFX	0	0.00
Hospital Sik	7	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	7	1.48
		DFO + DFP	0	0.00

		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO+ DFP + DFX	0	0.00
Hospital Baling	4	DFO only	0	0.00
		DFP only	3	0.64
		DFX only	1	0.21
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO+ DFP + DFX	0	0.00
		Total		

Table 5.10: 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	162	DFO only	11	6.79
		DFP only	16	9.88
		DFX only	130	80.25
		DFO + DFP	3	1.85
		DFP + DFX	1	0.62
		DFO + DFX	1	0.62
		DFO+ DFP + DFX	0	0.00
15–29.9	219	DFO only	33	15.07
		DFP only	66	30.14
		DFX only	69	31.51
		DFO + DFP	33	15.07
		DFP + DFX	9	4.11
		DFO + DFX	9	4.11
		DFO+ DFP + DFX	0	0.00
30–44.9	55	DFO only	1	1.82
		DFP only	37	67.27
		DFX only	3	5.45
		DFO + DFP	8	14.55
		DFP + DFX	1	1.82
		DFO + DFX	3	5.45
		DFO+ DFP + DFX	2	3.64
45–59.9	29	DFO only	5	17.24
		DFP only	14	48.28
		DFX only	2	6.90
		DFO + DFP	5	17.24

		DFP + DFX	1	3.45
		DFO + DFX	2	6.90
		DFO+ DFP + DFX	0	0.00
60 and above	7	DFO only	0	0.00
		DFP only	4	57.14
		DFX only	2	28.57
		DFO + DFP	1	14.29
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO+ DFP + DFX	0	0.00
		Total		

5.4.2 Serum Ferritin Level

Based on Table 5.11, 72 patients (22.29%) are with serum ferritin level lower than 1,000 ng/mL, 140 patients (43.34%) with serum ferritin level between 1,000-2,499 ng/mL, 65 patients (20.12%) with serum ferritin level between 2,500-4,999 ng/mL, and 46 patients (14.24%) with serum ferritin level above 5,000 ng/mL.

Table 5.11: Distribution of TDT 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 Sultanah Bahiyah	225	41	12.69	96	29.72	50	15.48	26	8.05	12	3.72
Hospital Sultan Abdul Halim	45	20	6.19	20	6.19	4	1.24	1	0.31	0	0.00
Hospital Kulim	40	8	2.48	19	5.88	9	2.79	2	0.62	2	0.62
Hospital Kuala Nerang	9	3	0.93	3	0.93	2	0.62	1	0.31	0	0.00
Hospital Sik	2	0	0.00	1	0.31	0	0.00	1	0.31	0	0.00
Hospital Baling	1	0	0.00	1	0.31	0	0.00	0	0.00	0	0.00
Hospital Sultanah Maliha	1	0	0.00	0	0.00	0	0.00	0	0.00	1	0.31
Total	323	72	22.29	140	43.34	65	20.12	31	9.60	15	4.64

5.5 COMPLICATIONS AND DEATHS

5.5.1 Complications

Based on Table 5.12, there was no new case of transfusion transmissible infection in 2020 among Thalassaemia patients in Kedah. This might mean safer blood are being given to patients.

Table 5.12: Distribution of Patients in Kedah According to Tranfusion Transmissible Infections

Complications	2007-2019	2020
Hepatitis C: Anti HCV HCV RNA	4	0
	0	0
Hepatitis B	0	0
HIV	0	0
Total	4	0

In Table 5.13, there are 386 TDT patients who are above 15 years old in Kedah. About 24 patients have delayed puberty, and 32 patients have short stature.

Table 5.13: Distribution of Patients in Kedah Accoding to Endocrine Complications

Complications	Number of Patients (n)
Short Stature	32
Delayed Puberty	24
Diabetes Mellitus	9
Hypothyroid	8
Total	73

5.5.2 Iron Deposition in Heart and Liver Complications

Tables 5.14 and 5.15 show that 274 patients had done liver and cardiac MRI T2* scans. Cardiac MRI T2* result shows that 243 (88.69%) cases iron content in the heart was in normal range, seven (2.55%) cases had mild cardiac iron deposition, 10 (3.65%) moderate and 14 (5.11%) cases have severe iron deposition. Liver MRI results shows that 19 (6.93%) cases with normal range iron content in the liver, 41 (14.96%) cases showed mild liver iron deposition, 71 (25.91%) cases are moderate and 143 (52.19%) cases with severe iron deposition.

Table 5.14: Distribution of Patients in Kedah According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Bahiyah	190	171	62.41	4	1.46	5	1.82	10	3.65
Hospital Sultan Abdul Halim	65	60	21.90	0	0.00	2	0.73	3	1.09
Hospital Kuala Nerang	15	10	3.65	2	0.73	2	0.73	1	0.36
Hospital Sik	3	1	0.36	1	0.36	1	0.36	0	0.00
Hospital Kulim	1	1	0.36	0	0.00	0	0.00	0	0.00
Total	274	243	88.69	7	2.55	10	3.65	14	5.11

Table 5.15: Distribution of Patients in Kedah According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Bahiyah	190	13	4.74	16	5.84	51	18.61	110	40.15
Hospital Sultan Abdul Halim	65	4	1.46	20	7.30	16	5.84	25	9.12
Hospital Kuala Nerang	15	0	0.00	4	1.46	4	1.46	7	2.55
Hospital Sik	3	1	0.36	1	0.36	0	0.00	1	0.36
Hospital Kulim	1	1	0.36	0	0.00	0	0.00	0	0.00
Total	274	19	6.93	41	14.96	71	25.91	143	52.19

5.5.3 Deaths Cases

As shown in Table 5.16, there were 42 deaths of thalassaemia patients in Kedah. The most common cause of death among thalassaemia patients was infection.

Table 5.16: Cumulative Known Causes of Death in Kedah

Causes of Death	Numbers of patients (n)
Infections	32
Motor Vehicle Accident (MVA)	3
Cardiac	2
Central Nervous System Event	2
Liver disease	1
Bone Marrow Transplant Complications	1
Others	1
Total	42

5.6 CONCLUSION

Kedah has a high number of thalassaemia patients in Malaysia. The commonest type of thalassaemia in Kedah is HbE/ β -Thalassaemia, with 366 patients (40.53%), followed by HbH Disease (300 patients, 33.22%) and 39 (4.32%) patients have other diagnoses. The median age group is 10-14.9 years old. A significant number of patients were lost to follow up and the majority are NTDT (164 patients, 17.6%).

Approximately 58% of the thalassaemia patients in Kedah is below 20 years old. A total of 472 patients are receiving chelation therapy, and oral DFX is the most common chelator used (43.64% of patients on chelators). Around 65% of patients achieved serum ferritin level below 2,500 ng/mL.

The most common endocrine complication occurred among pediatric patients is short stature. Based on 274 patients who had completed liver and cardiac MRI T2* scan, 11.31% have evidence of mild to severe cardiac iron loading and 52.19% have severe liver iron loading.

In Kedah, two hospitals that provide day care facilities are Hospital Sultan Abdul Halim (paediatric) and Hospital Sultanah Bahiyah (adult).

6.0

KELANTAN

6.1 INTRODUCTION

Kelantan is a state located in the northeast of Peninsular Malaysia. Kelantan’s population is estimated at 1.89 million in 2020. The population is dominated by those of Malay descent, followed by the Chinese, Siamese and Indians.

There are ten hospitals involved in providing treatment to thalassemia patients. These are Hospital Raja Perempuan Zainab II (HRPZII), Hospital Universiti Sains Malaysia (HUSM), Hospital Kuala Krai, Hospital Tanah Merah, Hospital Pasir Mas, Hospital Tengku Anis, Hospital Jeli, Hospital Machang, Hospital Tumpat and Hospital Gua Musang. HRPZII, HUSM and Hospital Kuala Krai are major specialist hospitals. Hospital Tanah Merah is a minor specialist hospital and others are non-specialist hospitals.

The total number of thalassemia patients in Kelantan registered in the registry in 2020 is 506 patients. More than half of these patients received treatment at Hospital Raja Perempuan Zainan II and Hospital Universiti Sains Malaysia. In Kelantan, HRPZII, HUSM and Hospital Kuala Krai provided day care services including blood transfusions for thalassemia patients.

6.2 PATIENT DEMOGRAPHICS

Table 6.1 shows the distribution of thalassemia patients by centre in Kelantan. HRPZII has the highest number of thalassemia patients whereas Hospital Tengku Anis has the least.

Table 6.1: Distribution of Patients in Kelantan by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Raja Perempuan Zainab II	227	44.86
Hospital Universiti Sains Malaysia	92	18.18
Hospital Kuala Krai	57	11.26
Hospital Tanah Merah	32	6.32
Hospital Gua Musang	27	5.34
Hospital Machang	21	4.15
Hospital Tumpat	19	3.75
Hospital Pasir Mas	15	2.96
Hospital Tengku Anis	8	1.58
Hospital Jeli	8	1.58
Total	506	100.00

Data shown included patients who are either living, lost to follow up or cured by bone marrow transplant. Total transfusion dependent (TDT) patients are 271 and 235 patients are non-transfusion dependent (NTDT) in Kelantan. To this point, there were only two patients successfully cured by bone marrow transplant (BMT) in Kelantan. In addition, no deaths of thalassaemia patients was recorded this year.

Table 6.2: Distribution of Patients in Kelantan by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	438
Cured by Stem Cell Therapy	2
Total	440
Lost to Follow- Up	66
Total	506
Death in 2020	0
Cumulative Reported Deaths	35

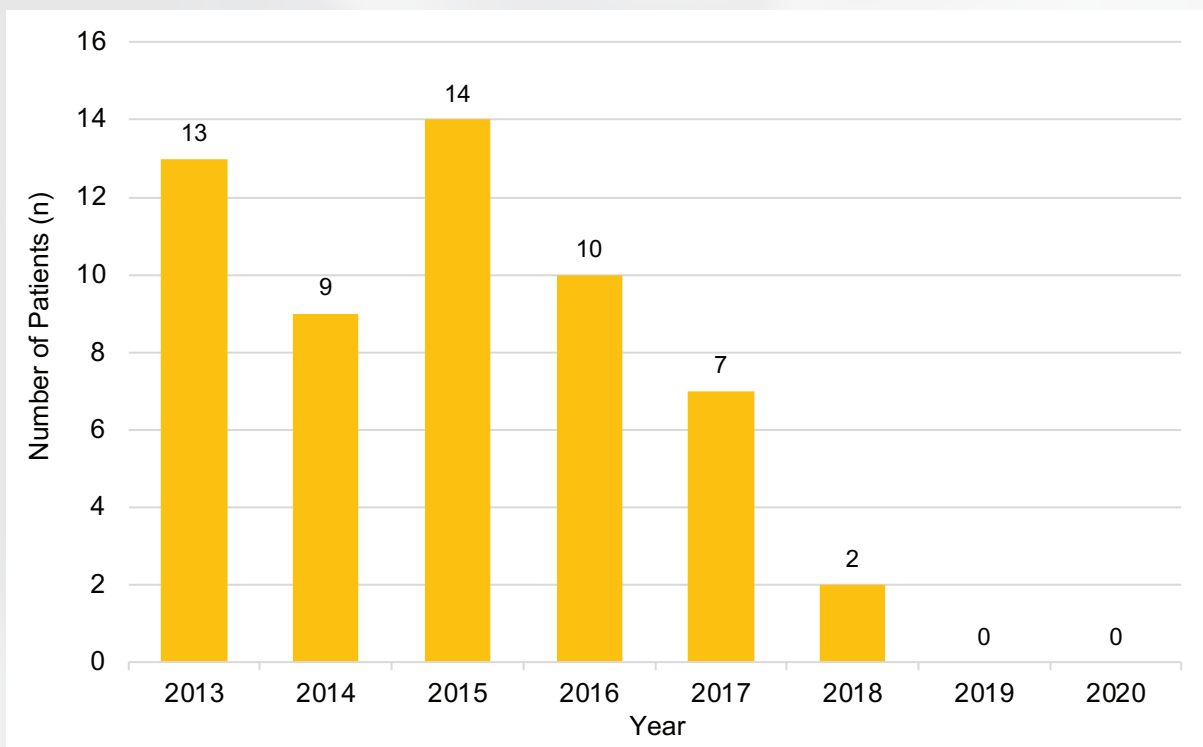


Figure 6.1: Distribution of Thalassaemia Births in Kelantan by Year

6.2.1 Age Groups

As shown in Figure 6.2, most patients in Kelantan diagnosed with thalassaemia were young adults. Patients aged between 20-24.9 years old lead with 98 patients (19.37%), followed by patients aged between 10-14.9 years old with 89 patients (17.59%). The age range with the least number of thalassaemia patients are 60-64.9 years old with two patients (0.40%).

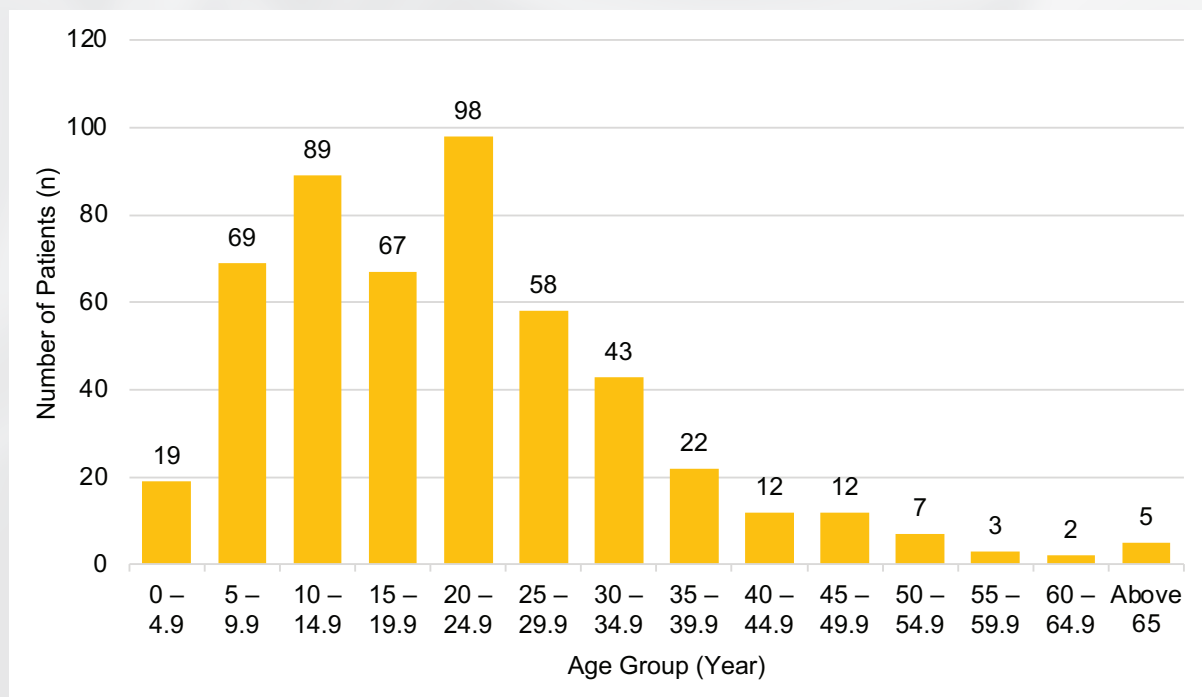


Figure 6.2: Distribution of Patients in Kelantan by Age Group

Table 6.3: Distribution of Patients in Kelantan According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0–14.9	177	β-Thalassemia Major	23	12.99
		β-Thalassemia Intermedia	15	8.47
		HbE/β-Thalassemia	83	46.89
		Hb H disease	46	25.99
		Others	10	5.65
15–29.9	223	β-Thalassemia Major	28	12.56
		β-Thalassemia Intermedia	4	1.79
		HbE/β-Thalassemia	135	60.54
		Hb H disease	52	23.32
		Others	4	1.79
30–44.9	77	β-Thalassemia Major	14	18.18
		β-Thalassemia Intermedia	7	9.09
		HbE/β-Thalassemia	35	45.45
		Hb H disease	20	25.97
		Others	1	1.30
45–59.9	22	β-Thalassemia Major	0	0.00
		β-Thalassemia Intermedia	7	31.82
		HbE/β-Thalassemia	8	36.36
		Hb H disease	4	18.18
		Others	3	13.64
60 and above	7	β-Thalassemia Major	0	0.00
		β-Thalassemia Intermedia	2	28.57
		HbE/β-Thalassemia	2	28.57
		Hb H disease	2	28.57
		Others	1	14.29
		Total	506	

6.2.2 Gender

The majority of the patients in Kelantan are female, about 270 patients (53.36%). Meanwhile, 236 (46.64%) of the patients are male.

Table 6.4: Distribution of Patients in Kelantan According to Gender by Centre

Centre	Total	Male		Female	
		No.	%	No.	%
Hospital Raja Perempuan Zainab II	227	109	21.54	118	23.32
Hospital Universiti Sains Malaysia	92	37	7.31	55	10.87
Hospital Kuala Krai	57	23	4.55	34	6.72
Hospital Tanah Merah	32	16	3.16	16	3.16
Hospital Gua Musang	27	15	2.96	12	2.37
Hospital Machang	21	8	1.58	13	2.57
Hospital Tumpat	19	8	1.58	11	2.17
Hospital Pasir Mas	15	7	1.38	8	1.58
Hospital Tengku Anis	8	7	1.38	1	0.20
Hospital Jeli	8	6	1.19	2	0.40
Total	506	236	46.64	270	53.36

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 due to its population is dominated by the Malays.

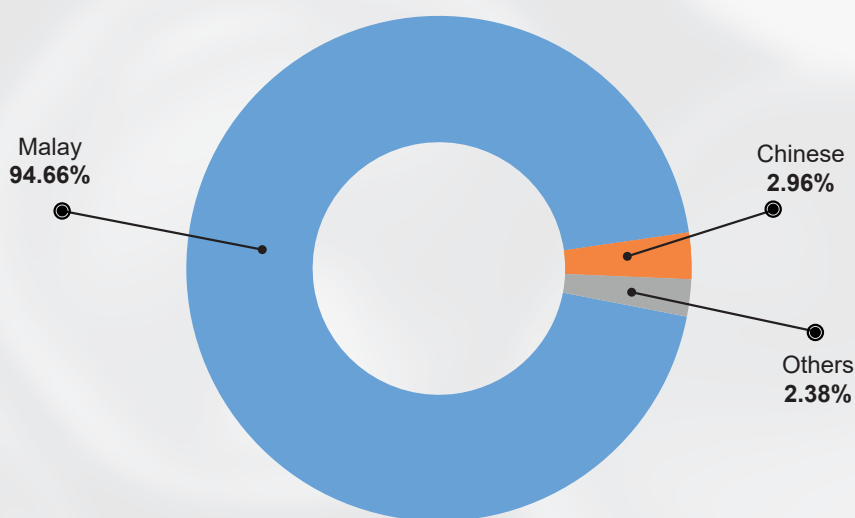


Figure 6.3: Distribution of Patients in Kelantan by Ethnic Group

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

Centre	Total Number of Patients	Malay		Chinese		Indian	
		No.	%	No.	%	No.	%
Hospital Raja Perempuan Zainab II	227	213	42.09	8	1.58	6	1.19
Hospital Universiti Sains Malaysia	92	86	17.00	6	1.19	0	0.00
Hospital Kuala Krai	57	57	11.26	0	0.00	0	0.00
Hospital Tanah Merah	32	32	6.32	0	0.00	0	0.00
Hospital Gua Musang	27	26	5.14	1	0.20	0	0.00
Hospital Machang	21	19	3.75	0	0.00	2	0.40
Hospital Tumpat	19	15	2.96	0	0.00	4	0.79
Hospital Pasir Mas	15	15	2.96	0	0.00	0	0.00
Hospital Tengku Anis	8	8	1.58	0	0.00	0	0.00
Hospital Jeli	8	8	1.58	0	0.00	0	0.00
Total	506	479	94.66	15	2.96	12	2.37

6.3 DIAGNOSIS

HbE/ β -thalassaemia is the main diagnosis of thalassaemia patients in Kelantan, followed by Hb H disease, β -thalassaemia major and β -thalassaemia intermedia. Another 19 patients were diagnosed with thalassaemia types other than those mentioned above.

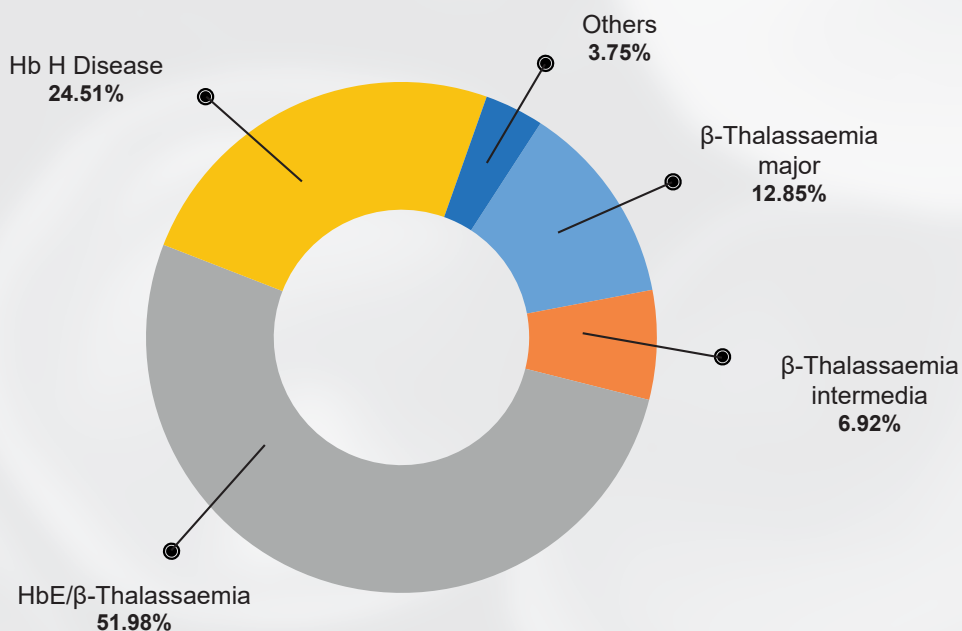


Figure 6.4: Distribution of Patients in Kelantan by Diagnosis

Based on Table 6.6, HRPZ II has the highest number of thalassaemia patients in each type of diagnosis compared to other hospitals as it is a tertiary hospital in Kelantan. Thus, most of the thalassaemia patients in Kelantan seeking treatments at HRPZ II.

Table 6.6: Distribution of Patients in Kelantan According to Diagnosis by Centre

Centre	Total Number of Patients	β-Thalassaemia major		β-Thalassaemia intermedia		HbE/β-Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Raja Perempuan Zainab II	227	20	3.95	18	3.56	123	24.31	59	11.66	7	1.38
Hospital Universiti Sains Malaysia	92	8	1.58	4	0.79	57	11.26	23	4.55	0	0.00
Hospital Kuala Krai	57	11	2.17	4	0.79	30	5.93	11	2.17	1	0.20
Hospital Tanah Merah	32	4	0.79	4	0.79	10	1.98	10	1.98	4	0.79
Hospital Gua Musang	27	6	1.19	1	0.20	15	2.96	5	0.99	0	0.00
Hospital Machang	21	3	0.59	2	0.40	9	1.78	6	1.19	1	0.20
Hospital Tumpat	19	3	0.59	2	0.40	6	1.19	5	0.99	3	0.59
Hospital Pasir Mas	15	6	1.19	0	0.00	4	0.79	3	0.59	2	0.40
Hospital Tengku Anis	8	2	0.40	0	0.00	5	0.99	1	0.20	0	0.00
Hospital Jeli	8	2	0.40	0	0.00	4	0.79	1	0.20	1	0.20
Total	506	65	12.85	35	6.92	263	51.98	124	24.51	19	3.75

Table 6.7 showed that Malay patients form the majority in all the four main types of thalassaemia in Kelantan.

Table 6.7: 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	65	Malay	62	12.25
		Chinese	2	0.40
		Others	1	0.20
β- Thalassaemia Intermedia	35	Malay	34	6.72
		Chinese	1	0.20
		Others	0	0.00
HbE/β-Thalassaemia	263	Malay	244	48.22
		Chinese	11	2.17
		Others	8	1.58
		Malay	121	23.91
		Chinese	0	0.00
Hb H Disease	124	Others	3	0.59
		Malay	18	3.56
		Chinese	1	0.20
Others	19	Others	0	0.00
		Chinese	0	0.00
		Indian	0	0.00
Total			506	100.00

6.4 TREATMENT

6.4.1 Iron Chelation

A total of 282 thalassaemia patients (55.73%) in Kelantan were prescribed chelating agents. DFX is the most commonly prescribed agent (prescribed to 128 patients out of 282, 45.39%), followed by DFP (67 patients, 23.76%) and DFO (44 patients, 15.60%).

Table 6.8: Distribution of Patients in Kelantan by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	44	15.60
DFP only	67	23.76
DFX only	128	45.39
DFO + DFP	15	5.32
DFP + DFX	12	4.26
DFO + DFX	15	5.32
DFO + DFP + DFX	1	0.35
Total	282	100.00

Table 6.9: 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	129	DFO only	11	3.90
		DFP only	35	12.41
		DFX only	63	22.34
		DFO + DFP	5	1.77
		DFP + DFX	5	1.77
		DFO + DFX	9	3.19
		DFO + DFP + DFX	1	0.35
Hospital Universiti Sains Malaysia	55	DFO only	20	7.09
		DFP only	13	4.61
		DFX only	7	2.48
		DFO + DFP	10	3.55
		DFP + DFX	1	0.35
		DFO + DFX	4	1.42
		DFO + DFP + DFX	0	0.00
Hospital Kuala Krai	33	DFO only	3	1.06
		DFP only	7	2.48
		DFX only	18	6.38
		DFO + DFP	1	0.35
		DFP + DFX	3	1.06
		DFO + DFX	1	0.35
		DFO + DFP + DFX	0	0.00
Hospital Tanah Merah	19	DFO only	0	0.00
		DFP only	4	1.42
		DFX only	15	5.32
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Gua Musang	18	DFO only	1	0.35
		DFP only	4	1.42
		DFX only	13	4.61
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Machang	9	DFO only	2	0.71
		DFP only	1	0.35
		DFX only	6	2.13
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Pasir Mas	6	DFO only	3	1.06
		DFP only	2	0.71
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	1	0.35
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tengku Anis	5	DFO only	3	1.06
		DFP only	0	0.00
		DFX only	2	0.71
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Tumpat	4	DFO only	0	0.00
		DFP only	1	0.35
		DFX only	3	1.06
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Jeli	4	DFO only	1	0.35
		DFP only	1	0.35
		DFX only	2	0.71
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			282	100.00

Table 6.10 indicates that iron chelators are mostly prescribed to patients below 30 years old. DFX was mainly prescribed to patients below 20 years old. So far, only one patient is prescribed with all three chelating agents.

Table 6.10: 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	91	DFO only	5	5.49
		DFP only	11	12.09
		DFX only	68	74.73
		DFO + DFP	1	1.10
		DFP + DFX	1	1.10
		DFO + DFX	5	5.49
		DFO + DFP + DFX	0	0.00
15–29.9	137	DFO only	32	23.36
		DFP only	35	25.55
		DFX only	50	36.50
		DFO + DFP	5	3.65
		DFP + DFX	7	5.11
		DFO + DFX	7	5.11
		DFO + DFP + DFX	1	0.73
30–44.9	39	DFO only	5	12.82
		DFP only	14	35.90
		DFX only	9	23.08
		DFO + DFP	7	17.95
		DFP + DFX	2	5.13
		DFO + DFX	2	5.13
		DFO + DFP + DFX	0	0.00
45–59.9	12	DFO only	1	8.33
		DFP only	6	50.00
		DFX only	2	16.67
		DFO + DFP	3	25.00
		DFP + DFX	0	0.00
		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			282	

6.4.2 Serum Ferritin Level

Based on Table 6.11, there are 70 TDT patients in Kelantan who have their serum ferritin level regularly updated. A total of 37 patients have serum ferritin level lower than 2,500 ng/mL.

Table 6.11: Distribution of TDT 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	28	3	4.29	15	21.43	9	12.86	1	1.43	0	0.00
Hospital Kuala Krai	23	4	5.71	7	10.00	6	8.57	5	7.14	1	1.43
Hospital Universiti Sains Malaysia	8	2	2.86	1	1.43	3	4.29	1	1.43	1	1.43
Hospital Machang	7	1	1.43	1	1.43	2	2.86	2	2.86	1	1.43
Hospital Gua Musang	3	1	1.43	1	1.43	1	1.43	0	0.00	0	0.00
Hospital Tumpat	1	0	0.00	1	1.43	0	0.00	0	0.00	0	0.00
Total	70	11	15.71	26	37.14	21	30.00	9	12.86	3	4.29

6.5 COMPLICATIONS AND DEATHS

6.5.1 Complications

The data about complications of patients for Kelantan could not be translated due to lack of information.

6.5.2 Iron Deposition in Heart and Liver Complications

Table 6.12 and 6.13 shows that only two patients had their liver and cardiac MRI T2* result.

Table 6.12: Distribution of Patients in Kelantan According to Cardiac MRI T2*

Grade of Iron Deposition	Number of Patients (n)	Percentage (%)
Severe (<10)	0	0.00
Moderate (10-15)	0	0.00
Mild (15-20)	1	50.00
Normal (>20)	1	50.00
Total	2	100.00

Table 6.13: Distribution of Patients in Kelantan According to Liver MRI

Grade of Iron Deposition	Number of Patients (n)	Percentage (%)
Severe (<1.8)	0	0.00
Moderate (1.8-3.8)	1	50.00
Mild (3.8-11.4)	1	50.00
Normal (>11.4)	0	0.00
Total	2	100.00

6.5.3 Death Cases

Table 6.14 shows that there have been 33 cumulative known causes of death for thalassemia patients in Kelantan, excluding two cases of unknown cause of death due to the patients who died at home. The most common causes of death among thalassemia patients were infections and cardiac.

Table 6.14: Cumulative Known Cause of Death in Kelantan

Causes Of Death	Number of Patients (n)
Cardiac	15
Infections	14
Endocrine Complication	1
Thrombosis	1
Died at Home/Brought in Dead to Hospital	1
Others	1
Total	33

6.6 CONCLUSION

The total number of thalassemia patients in Kelantan is 506. The majority of thalassemia patients receive treatment at tertiary centres, such as HRPZ II (227 patients out of 506, 44.86%) and HUSM (92 patients out of 506, 18.18%). In 2020, no death of thalassemia patients recorded in the state.

Currently, only two patients were reported as cured by stem cell transplant. The male and female patient distribution slightly differs at 236 (46.64%) for male and 270 patients (53.36%) for female. Most of the thalassemia patients in Kelantan are Malays (479 patients, 94.66%). HbE/ β -thalassemia contributed the greatest number of patients (263 patients, 51.98%), followed by Hb H disease (124 patients, 24.51%) and β -thalassemia major (65 patients, 12.85%).

There are 282 patients who are prescribed with iron chelators in Kelantan. The report shows that DFX is the most prescribed iron chelator to 128 patients (45.39%), followed by DFP to 67 patients (23.76%) and DFO to 44 patients (15.60%).

There are 70 (25.8%) out of 271 TDT patients who had their serum ferritin levels recorded and updated in their notes. About 37 patients of the 271 TDT patients have a serum ferritin level lower than 2,500 ng/mL. The remaining 33 patients have a serum ferritin level higher than 2,500 ng/mL.

Blood transfusion is the main treatment for thalassaemia patients. However, this treatment may also lead to other complications either directly or indirectly. Due to lack of information, complications for thalassaemia patients in Kelantan cannot be translated well. In 2020, only two patients had done liver and cardiac MRI T2* scan.



7.0
MELAKA

7.1 INTRODUCTION

Melaka was known as the Historical State. There are 3 districts in Melaka: Melaka Tengah, Alor Gajah and Jasin. The capital is Melaka Town.

Melaka has a population of 932,700 as of 2020, comprising of Bumiputera (617,400, 66.19%), Chinese (including the Peranakan community; 217,600, 23.33%), Indian (including the Chitty community; 51,700, 5.54%), other races (6,000, 0.64%), and non-Malaysian citizens (40,000, 4.29%) according to Department of Statistics Malaysia.

There are three government hospitals in the state of Melaka. Hospital Alor Gajah and Hospital Jasin are without specialist and all thalassaemia patients are referred to and managed by Hospital Melaka.

7.2 PATIENT DEMOGRAPHICS

Based on Table 7.1, there are 256 living thalassaemia patients in Melaka. An additional 10 patients compared to 2019. Four patients in Melaka have done bone marrow transplant within this year. There were two deaths in 2020 while a total of three patients died in 2019. However, only two deaths were documented in the 2019 annual report.

Table 7.1: Distribution of Patients in Melaka by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Melaka	256	100.00
Total	256	100.00

Table 7.2: Distribution of Patients in Melaka by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	219
Cured by Stem Cell Therapy	9
Total	228
Lost to Follow-up	28
Total	256
Death in 2020	2
Cumulative Reported Death	8

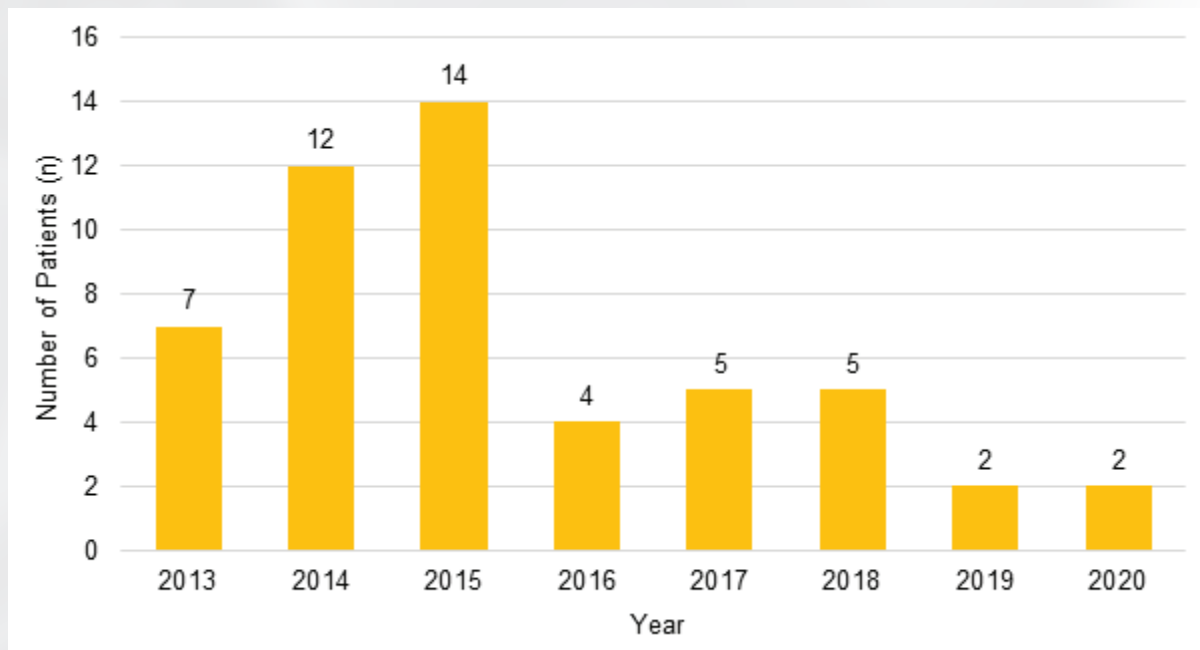


Figure 7.1: Distribution of Thalassaemia Births in Melaka by Year

7.2.1 Age Group

The youngest patient in Melaka is one year old and the oldest is 66 years old. The age groups 5-9.9 and 10-14.9 represent 40.2% of thalassaemia patients in Melaka. As shown in Table 7.3, HbE/ β -thalassaemia is the most common diagnosis for patients below 60 years old. The two oldest patients are diagnosed with Hb H disease.

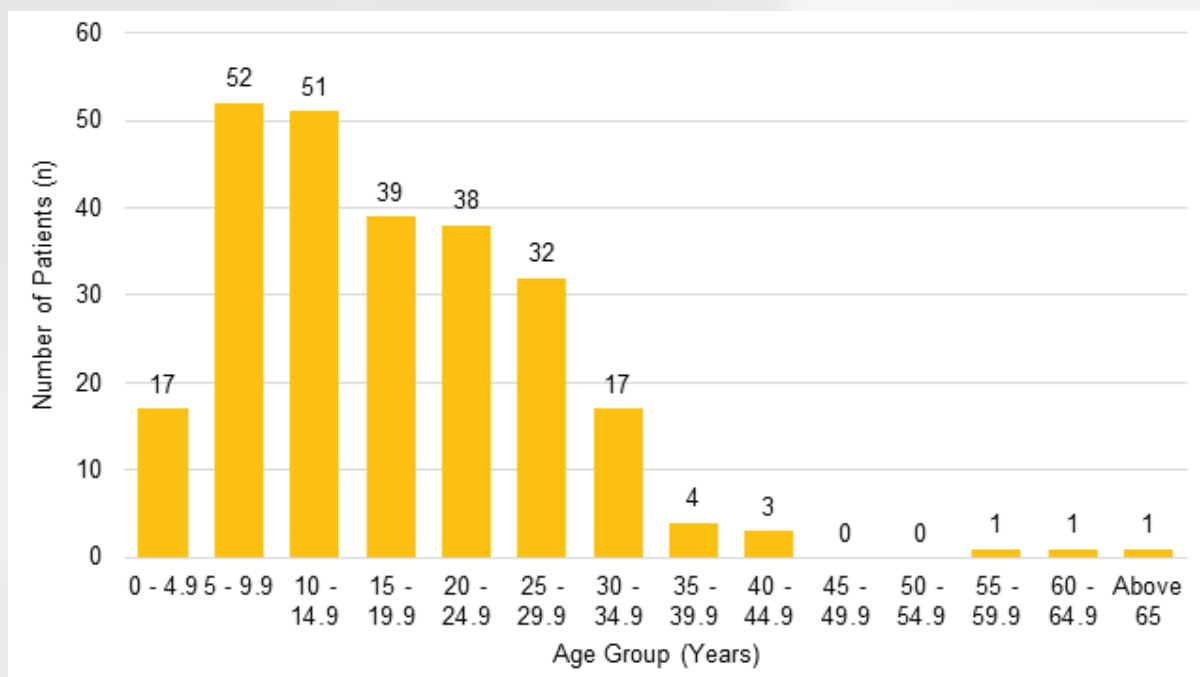


Figure 7.2: Distribution of Patients in Melaka by Age Group

Table 7.3: Distribution of Patients in Melaka According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0-14.9	120	β-Thalassaemia Major	24	20.00
		β-Thalassaemia Intermedia	2	1.67
		HbE/β-Thalassaemia	46	38.33
		Hb H Disease	39	32.50
		Others	9	7.50
15-29.9	109	β-Thalassaemia Major	27	24.77
		β-Thalassaemia Intermedia	3	2.75
		HbE/β-Thalassaemia	52	47.71
		Hb H Disease	24	22.02
		Others	3	2.75
30-44.9	24	β-Thalassaemia Major	4	16.67
		β-Thalassaemia Intermedia	4	16.67
		HbE/β-Thalassaemia	10	41.67
		Hb H Disease	6	25.00
		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
		Hb H Disease	0	0.00
		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
		Hb H Disease	2	100.00
		Others	0	0.00
		Total	256	

7.2.2 Gender

Table 7.4 shows the distribution of thalassaemia patients in Melaka by gender. There are 125 (48.83%) male patients and 131 (51.17%) female patients in Melaka.

Table 7.4: Distribution of Patients in Melaka According to Gender

Centre	Total Number of Patients	Male		Female	
		No.	%	No.	%
Hospital Melaka	256	125	48.83	131	51.17
Total	256	125	48.83	131	51.17

7.2.3 Ethnic Group

Thalassaemia patients in Melaka are mostly Malays (205 patients), followed by Chinese (43 patients) and Indian (3 patients). Patients of other ethnicities include Bajau (1 patient), Iban (1 patient), Thais (2 patients) and Pribumi Sabah (1 patient).

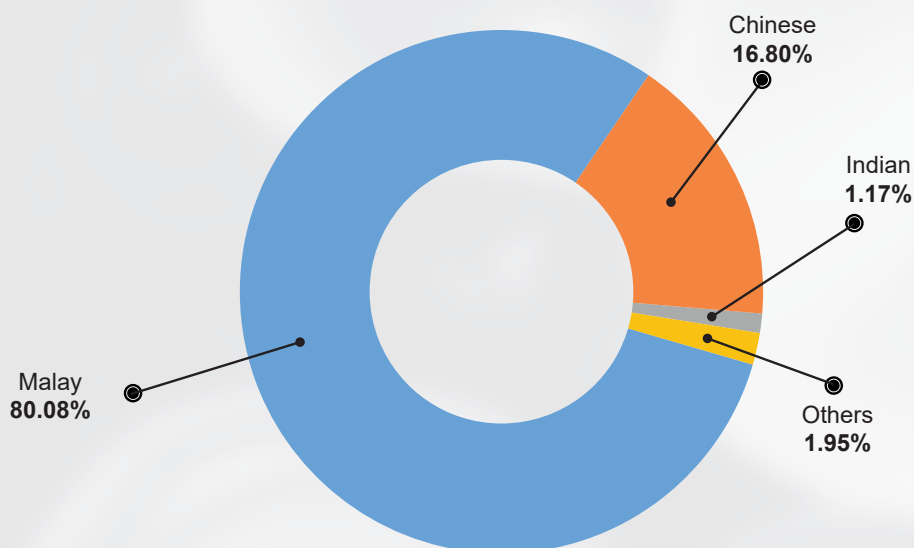


Figure 7.3: Distribution of Patients in Melaka by Ethnic Group

7.3 DIAGNOSIS

As shown in Figure 7.4, HbE/ β -thalassaemia is the main diagnosis with 109 patients (42.58%), followed by Hb H disease with 71 patients (27.73%), β -thalassaemia major with 55 patients (21.48%) and β -thalassaemia intermedia with only nine patients (3.52%). From Table 7.5, Malay patients are most commonly diagnosed with HbE/ β -thalassaemia, whereas the majority of Chinese patients are diagnosed with β -thalassaemia major.

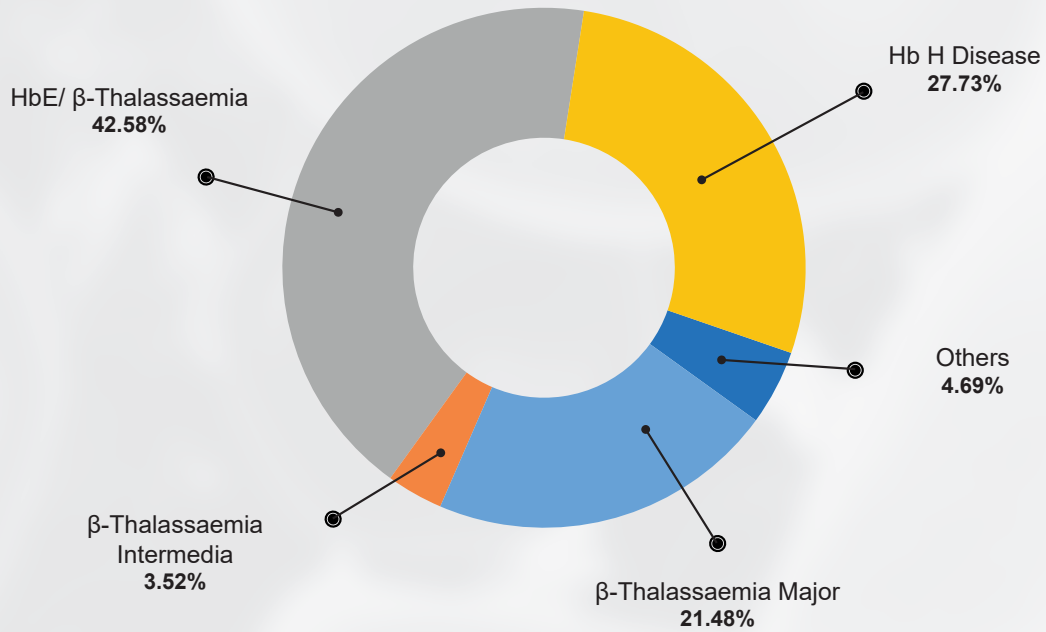


Figure 7.4: Distribution of Patients in Melaka by Diagnosis

Table 7.5: 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	55	Malay	35	13.67
		Chinese	20	7.81
		Indian	0	0.00
		Kadazan Dusun	0	0.00
		Others	0	0.00
β-Thalassaemia Intermedia	9	Malay	6	2.34
		Chinese	2	0.78
		Indian	0	0.00
		Kadazan Dusun	0	0.00
		Others	1	0.39
HbE/β-Thalassaemia	109	Malay	102	39.84
		Chinese	4	1.56
		Indian	2	0.78
		Kadazan Dusun	0	0.00
		Others	1	0.39

Hb H Disease	71	Malay	51	19.92
		Chinese	17	6.64
		Indian	0	0.00
		Kadazan Dusun	0	0.00
		Others	3	1.17
Others	12	Malay	11	4.30
		Chinese	0	0.00
		Indian	1	0.39
		Kadazan Dusun	0	0.00
		Others	0	0.00
Total			256	100.00

7.4 TREATMENT

7.4.1 Iron Chelation Therapy

Table 7.6 shows that there are 137 patients who receive iron chelators in Melaka (53.52%). Of these, 34 patients (24.82%) are on DFO, 27 patients (19.71%) are on DFP, 48 patients (35.04%) are on DFX and 28 patients (20.44%) receive a combination of two iron chelators. The youngest patient in Melaka receiving iron chelation therapy is four years old. As shown in Table 7.7, the younger cohort of patients from age group 0-14.9 years old mostly prescribed DFX. Patients from age group 15-59.9 years old who are on iron chelators commonly receive DFO monotherapy. There are two patients above 60 years old who are given DFP monotherapy.

Table 7.6: Distribution of Patients in Melaka by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	34	24.82
DFP only	27	19.71
DFX only	48	35.04
DFO + DFP	17	12.41
DFP + DFX	8	5.84
DFO + DFX	3	2.19
DFO + DFP + DFX	0	0.00
Total	137	100.00

Table 7.7: 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	48	DFO only	8	16.67
		DFP only	2	4.17
		DFX only	37	77.08
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	2.08
		DFO + DFP + DFX	0	0.00
15-29.9	69	DFO only	20	28.99
		DFP only	14	20.29
		DFX only	11	15.94
		DFO + DFP	16	23.19
		DFP + DFX	7	10.14
		DFO + DFX	1	1.45
		DFO + DFP + DFX	0	0.00
30-44.9	17	DFO only	6	35.29
		DFP only	9	52.94
		DFX only	0	0.00
		DFO + DFP	1	5.88
		DFP + DFX	0	0.00
		DFO + DFX	1	5.88
		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	0	0.00
		DFP + DFX	1	100.00
		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
Total			137	

7.4.2 Serum Ferritin Level

There are 102 TDT patients who had their serum ferritin level measured. According to Table 7.8, there are two patients had serum ferritin level more than 10,000 ng/mL. The highest serum ferritin level measured was 11,891.30 ng/mL (10 years old patient with HbE/ β -thalassaemia). More than half of TDT patients (61.76%) have serum ferritin levels lower than 2,499 ng/mL

Table 7.8: Distribution of TDT Patients in Melaka According to Most 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 Melaka	102	23	22.55	40	39.22	24	23.53	13	12.75	2	1.96
Total	102	23	22.55	40	39.22	24	23.53	13	12.75	2	1.96

7.5 COMPLICATIONS AND DEATHS

7.5.1 Complications

As shown in Table 7.9, there is one patient that reported with tranfusion transmissible infection of Hepatitis C. Nevertheless, there was no new tranfusion transmissible infection in 2020.

Table 7.9: Distribution of Patients in Melaka According to Tranfusion Transmissible Infections

Complications	2007-2019	2020
Hepatitis C: Anti HCV HCV RNA	1	0
	0	0
Hepatitis B	0	0
HIV	0	0
Total	1	0

Based on the Table 7.10, there were 20 patients who had complications, such as short stature with eight patients, followed by delayed puberty with seven patients, three patients with hypothyroid and two patients with diabetes mellitus.

Table 7.10: Distribution of Patients in Melaka According to Endocrine Complications

Complications	Number of Patients (n)
Short Stature	8
Delayed Puberty	7
Hypothyroid	3
Diabetes Mellitus	2
Total	20

7.5.2 Iron Deposition in Heart and Liver Complications

A total of 33 thalassaemia patients in Melaka had undergone liver and cardiac MRI T2* scan. Based on Table 7.11, 27 (81.82%) patients had normal myocardial iron loading and two (6.06%) had severe myocardial iron loading. In contrast, 15 patients (45.45%) had severe iron loading in the liver and only three patients (9.09%) had normal iron loading.

Table 7.11: Distribution of Patients in Melaka According to Cardiac MRI T2*

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Melaka	33	27	81.82	2	6.06	2	6.06	2	6.06
Total	33	27	81.82	2	6.06	2	6.06	2	6.06

Table 7.12: Distribution of Patients in Melaka According to Liver MRI

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Melaka	33	3	9.09	8	24.24	7	21.21	15	45.45
Total	33	3	9.09	8	24.24	7	21.21	15	45.45

7.5.3 Deaths Cases

A total of eight deceased thalassaemia patients were reported in Melaka. The causes of death were multiple, such as cardiac, endocrine, infections and malignancy. There were two deaths recorded in year 2020 due to cardiac complication. Based on Table 7.13, the most common cause of death is cardiac.

Table 7.13: Cumulative Known Cause of Death in Melaka

Causes of Death	Number of Patients (n)
Cardiac	4
Infections	2
Endocrine complications	1
Malignancy	1
Total	8

7.6 CONCLUSION

There are 256 living patients currently registered at Hospital Melaka. Of these, 219 are active patients, nine patients are cured by stem cell therapy and 28 patients are lost to follow up. There is 1 thalassaemia birth reported in 2020. Age group 5-9.9 and 10-14.9 represent 40.2% of thalassaemia patients in Melaka. The oldest surviving β -thalassaemia major patients is in the age range of 30-34.9 years old.

Most of the TDT patients in Melaka were diagnosed with HbE/ β -thalassaemia. On the other hand, the highest number of NTDT patients were diagnosed as HbH disease.

A total of 74.45% of the 137 patients receiving regular transfusion have their serum ferritin level measured. Of these, 61.76% have a serum ferritin level below 2,499 ng/mL and 38.24% have a serum ferritin level above 2,500 ng/mL. Short stature and delayed puberty are the most common endocrine complications in Melaka. Out of 33 patients who had liver and cardiac MRI T2* done, 81.82% patients have normal myocardial iron loading. Meanwhile, 45.45% patients had severe liver iron loading. Since 2007, eight deaths were recorded in the registry. The main cause of death was cardiac.

8.0

**NEGERI
SEMBILAN**

8.1 INTRODUCTION

Negeri Sembilan is located on the western coast of Peninsular Malaysia. This state consists of seven districts, namely, Seremban, Port Dickson, Kuala Pilah, Jempol, Jelebu, Rembau and Tampin. The capital of Negeri Sembilan is Seremban.

Based on the Department of Statistics Malaysia, Negeri Sembilan has a collective population of 1.13 million in 2020 with the Malay community making up the majority of 59.2%, followed by Chinese 20.32%, Indians 13.64% and other ethnicities making up the remaining 6.84%.

There are seven government hospitals in Negeri Sembilan namely Hospital Tuanku Ja'afar, Hospital Tuanku Ampuan Najihah, Hospital Jelebu, Hospital Port Dickson, Hospital Jempol, Hospital Rembau and Hospital Tampin. However, there are no thalassaemia cases registered in the MTR from Hospital Jempol, Hospital Rembau and Hospital Tampin. Most of the thalassaemia patients receive their treatment at Hospital Tuanku Ja'afar, Seremban as it also provides day care services for blood transfusions.

8.2 PATIENT DEMOGRAPHICS

The total number of living patients registered up to 31st December 2020 in MTR is 233 patients. There was no new death reported in 2020.

Table 8.1: Distribution of Patients in Negeri Sembilan by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Tuanku Ja'afar	186	79.83
Hospital Tuanku Ampuan Najihah	41	17.60
Hospital Port Dickson	3	1.29
Hospital Jelebu	3	1.29
Total	233	100.00

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

Vital Status	Number of Patients (n)
Alive and On Active Treatment	194
Cured by Stem Cell Therapy	10
Total	204
Lost to Follow Up	29
Total	233
Death in 2020	0
Cumulative Reported Death	13

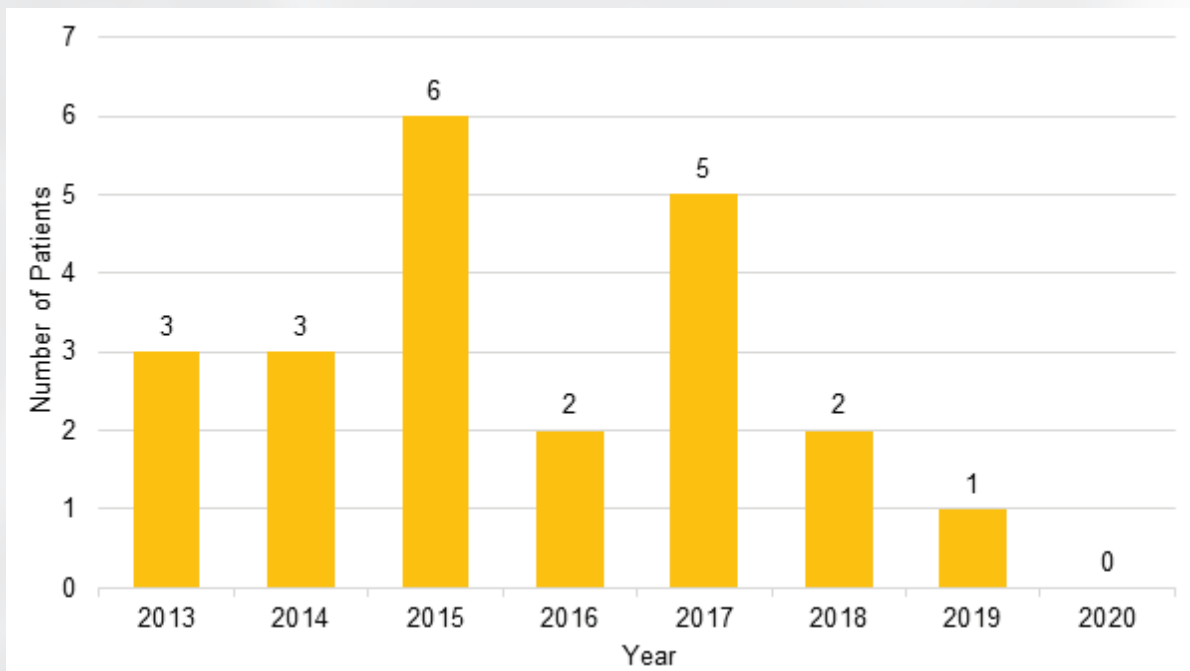


Figure 8.1: Distribution of Thalassaemia Births in Negeri Sembilan by Year

8.2.1 Age Groups

The youngest thalassaemia patient in Negeri Sembilan is two years old while the oldest patient is 68 years old. Based on Table 8.3, the highest number of patients are in the age group of 20-24.9 years old.

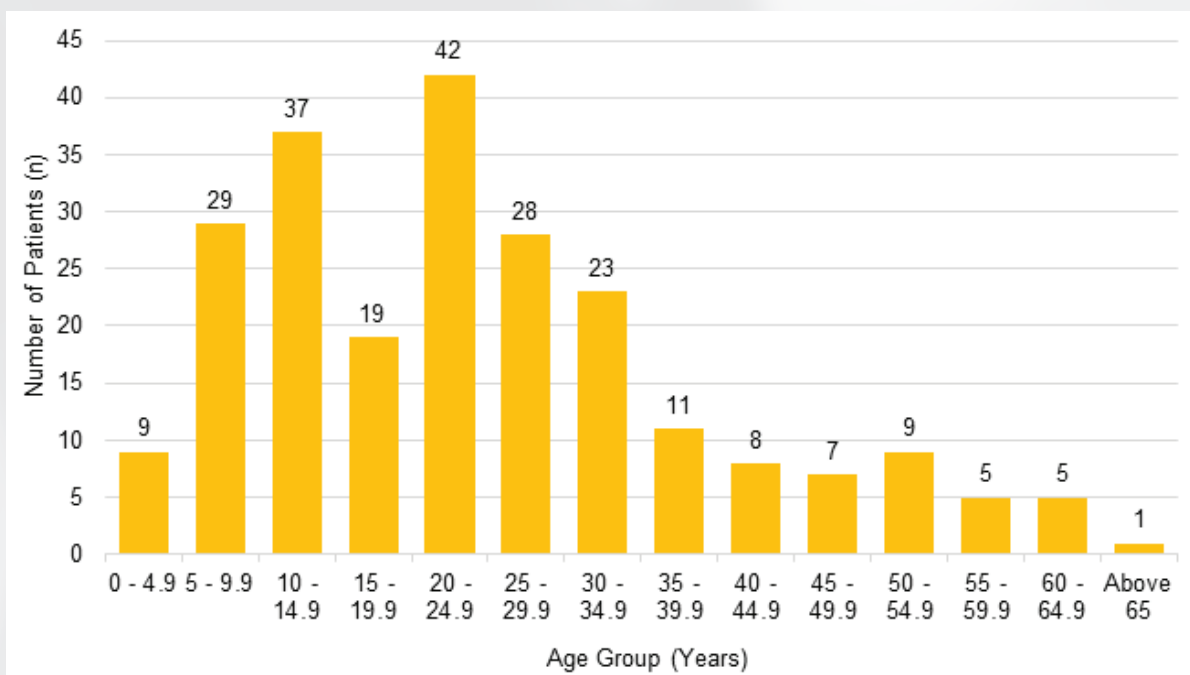


Figure 8.2: Distribution of Patients in Negeri Sembilan by Age Group

Table 8.3: Distribution of Patients in Negeri Sembilan According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0-14.9	75	β -Thalassaemia Major	18	24.00
		β -Thalassaemia Intermedia	5	6.67
		HbE/ β -Thalassaemia	28	37.33
		Hb H disease	24	32.00
		Others	0	0.00
15-29.9	89	β -Thalassaemia Major	26	29.21
		β -Thalassaemia Intermedia	3	3.37
		HbE/ β -Thalassaemia	35	39.33
		Hb H disease	20	22.47
		Others	5	5.62
30-44.9	42	β -Thalassaemia Major	7	16.67
		β -Thalassaemia Intermedia	6	14.29
		HbE/ β -Thalassaemia	13	30.95
		Hb H disease	16	38.10
		Others	0	0.00
45-59.9	21	β -Thalassaemia Major	0	0.00
		β -Thalassaemia Intermedia	7	33.33
		HbE/ β -Thalassaemia	5	23.81
		Hb H disease	9	42.86
		Others	0	0.00
60 and above	6	β -Thalassaemia Major	0	0.00
		β -Thalassaemia Intermedia	3	50.00
		HbE/ β -Thalassaemia	2	33.33
		Hb H disease	1	16.67
		Others	0	0.00
Total			233	

8.2.2 Gender

Table 8.4 shows that majority of the Thalassaemia patients in Negeri Sembilan are female with 144 patients (61.80%) and the remaining 89 patients (38.20%) are male.

Table 8.4: Distribution of Patients in Negeri Sembilan According to Gender by Centre

Centre	Total Number of Patients	Male		Female	
		No.	%	No.	%
Hospital Tuanku Ja'afar	186	62	26.61	124	53.22
Hospital Tuanku Ampuan Najihah	41	24	10.30	17	7.30
Hospital Jelebu	3	2	0.86	1	0.43
Hospital Port Dickson	3	1	0.43	2	0.86
Total	233	89	38.20	144	61.80

8.2.3 Ethnic Group

Figure 8.3 illustrates the ethnic group with the highest number of thalassaemia patients in Negeri Sembilan is Malay with 189 patients (81.12%), followed by Chinese with 35 patients (15.02%) and Indian with one patient (0.43%). The other ethnicities are Orang Asli with four patients (1.72%) and Kadazan Dusun with two patients (0.86%). There are also two foreigners treated as thalassaemia patients in Negeri Sembilan. One patient is from Indonesia and one patient is from Thailand.

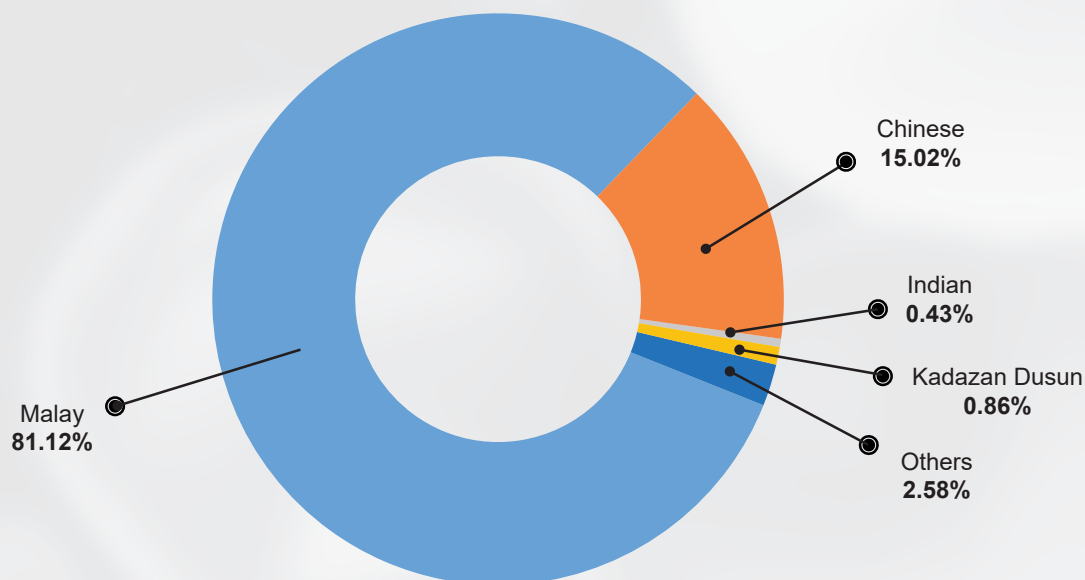


Figure 8.3: Distribution of Patients in Negeri Sembilan by Ethnic Groups

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

Centre	Total Number of Patients	Malay		Chinese		Indian		Kadazan Dusun		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Tuanku Ja'afar	186	151	64.81	31	13.30	1	0.43	2	0.86	1	0.43
Hospital Tuanku Ampuan Najihah	41	33	14.16	4	1.72	0	0.00	0	0.00	4	1.72
Hospital Jelebu	3	3	1.29	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Port Dickson	3	2	0.86	0	0.00	0	0.00	0	0.00	1	0.43
Total	233	189	81.12	35	15.02	1	0.43	2	0.86	6	2.58

8.3 DIAGNOSIS

The majority of patients in Negeri Sembilan were diagnosed with HbE/ β -thalassaemia (83 patients), followed by Hb H disease (70 patients), β -thalassaemia major (51 patients), β -thalassaemia intermedia (24 patients) and five patients were from other diagnosis.

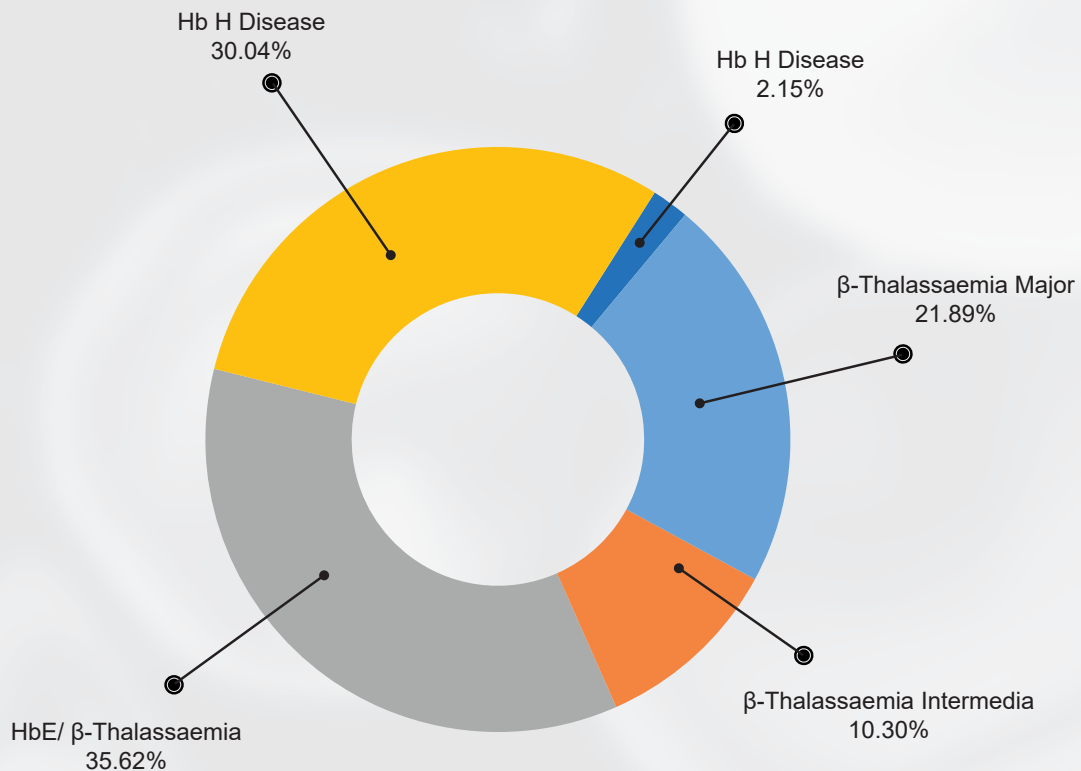


Figure 8.4: Distribution of Patients in Negeri Sembilan by Diagnosis

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

Centre	Total Number of Patients	β -Thalassaemia major		β -Thalassaemia intermedia		HbE/ β -Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Tuanku Ja'afar	186	43	18.45	24	10.30	68	29.18	50	21.46	1	0.43
Hospital Tuanku Ampuan Najihah	41	5	2.15	0	0.00	12	5.15	20	8.58	4	1.72
Hospital Jelebu	3	3	1.29	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Port Dickson	3	0	0.00	0	0.00	3	1.29	0	0.00	0	0.00
Total	233	51	21.89	24	10.30	83	35.62	70	30.04	5	2.15

Table 8.7: 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	51	Malay	34	14.59
		Chinese	15	6.44
		Indian	0	0.00
		Kadazan-Dusun	2	0.86
		Others	0	0.00
β-Thalassaemia Intermedia	24	Malay	20	8.58
		Chinese	4	1.72
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
HbE/β-Thalassaemia	83	Malay	79	33.91
		Chinese	1	0.43
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	3	1.29
Hb H Disease	70	Malay	53	22.75
		Chinese	13	5.58
		Indian	1	0.43
		Kadazan-Dusun	0	0.00
		Others	3	1.29
Others	5	Malay	3	1.29
		Chinese	2	0.86
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Total			233	100.00

8.4 TREATMENT

8.4.1 Iron Chelation Therapy

Based on Table 8.8, there are 129 patients (55.36%) that are prescribed iron chelation therapy. Out of this, 54 patients (41.86%) are on DFP, 36 patients (27.91%) are on DFX and 12 patients (9.30%) are on DFO. The remaining 27 patients (20.94%) are on combination of 2 iron chelators.

Out of 233 total patients in this state, there are 103 patients (44.21%) who are classified as TDT patients and the remaining 130 patients (55.79%) are classified as NTD patients. Based on Table 8.9, the majority of patients that received iron chelation therapy are those who are regularly transfused. It is mainly due to reduce iron overload accumulating in the body from the blood transfusions they received.

From a total of 103 TDT patients, the majority, 102 patients (99.03%) are on iron chelation therapy and only one TDT patient who is not prescribed any iron chelator, a foreigner that could not manage to pay the chelator. For NTD T patients, only 27 patients (20.77%) out of 130 patients are on iron chelation therapy.

Table 8.10 shows that most patients on chelation therapy are received treatment at Hospital Tuanku Ja'afar. Moreover, Table 8.11 shows that most of the paediatric patients (under 15 years old) are on DFX monotherapy. The majority of the patients that required combination iron chelators are adults.

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

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	12	9.30
DFP only	54	41.86
DFX only	36	27.91
DFO + DFP	17	13.18
DFP + DFX	1	0.78
DFO + DFX	9	6.98
DFO + DFP + DFX	0	0.00
Total	129	100.00

Table 8.9: Distribution of Patients in Negeri Sembilan According to Iron Chelation Therapy Status by Transfusion Status

Transfusion Status	Total Number of Patients	With Iron Chelation Therapy	Percentage (%)	Without Iron Chelation Therapy	Percentage (%)
TDT	103	102	99.03	1	0.97
NTDT	130	27	20.77	103	79.23
Total	233	129	55.36	104	44.64

Table 8.10: 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 Tuanku Ja'afar	112	DFO only	10	7.75
		DFP only	49	37.98
		DFX only	28	21.71
		DFO + DFP	16	12.40
		DFP + DFX	1	0.78
		DFO + DFX	8	6.20
		DFO + DFP + DFX	0	0.00
Hospital Tuanku Ampuan Najihah	13	DFO only	1	0.78
		DFP only	5	3.88
		DFX only	6	4.65
		DFO + DFP	1	0.78
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Port Dickson	2	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	2	1.55
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Jelebu	2	DFO only	1	0.78
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.78
		DFO + DFP + DFX	0	0.00
Total			129	100.00

Table 8.11: 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	38	DFO only	3	7.89
		DFP only	0	0.00
		DFX only	31	81.58
		DFO + DFP	2	5.26
		DFP + DFX	0	0.00
		DFO + DFX	2	5.26
		DFO + DFP + DFX	0	0.00
15.0-29.9	57	DFO only	7	12.28
		DFP only	25	43.86
		DFX only	4	7.02
		DFO + DFP	13	22.81
		DFP + DFX	1	1.75
		DFO + DFX	7	12.28
		DFO + DFP + DFX	0	0.00
30.0-44.9	16	DFO only	0	0.00
		DFP only	13	81.25
		DFX only	1	6.25
		DFO + DFP	2	12.50
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45.0-59.9	16	DFO only	2	12.50
		DFP only	14	87.50
		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	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
Total			129	

8.4.2 Serum Ferritin Level

Table 8.12 indicates there are only 96 out of 103 TDT patients in Negeri Sembilan had their serum ferritin measured in 2020. The lowest ferritin level recorded is 357.80 ng/mL and the highest ferritin level recorded is 19 677.04 ng/mL. As a result, 53.13% of patients had serum ferritin level less than 2,499 ng/mL.

Table 8.12: Distribution of TDT Patients in Negeri Sembilan According to Most 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 Tuanku Ja'afar	81	11	11.46	35	36.46	21	21.88	11	11.46	3	3.13
Hospital Tuanku Ampuan Najihah	11	0	0.00	2	2.08	5	5.21	3	3.13	1	1.04
Hospital Port Dickson	3	0	0.00	2	2.08	1	1.04	0	0.00	0	0.00
Hospital Jelebu	1	0	0.00	1	1.04	0	0.00	0	0.00	0	0.00
Total	96	11	11.46	40	41.67	27	28.13	14	14.58	4	4.17

8.5 COMPLICATIONS AND DEATHS

8.5.1 Complications

From Table 8.13, there are five patients from Hospital Tunku Ja'afar that are reported with transfusion transmissible infections of Hepatitis B and C. However, there is no new case of transfusion transmissible infection reported in Negeri Sembilan in 2020.

Table 8.13: Distribution of Patients in Negeri Sembilan According to Transfusion Transmissible Infections

Complications	2007-2019	2020
Hepatitis C: Anti HCV HCV RNA	4	0
	0	0
Hepatitis B	1	0
HIV	0	0
Total	5	0

Table 8.14 shows 22 patients had endocrine complications, namely, two patients had short stature, two female patients with delayed puberty / hypogonadism, eight patients from the ages of 35 to 61 years old with diabetes mellitus and 10 patients had hypothyroid.

Table 8.14: Distribution of Patients in Negeri Sembilan According to Endocrine Complications by Centre

Complications	Number of Patients (n)
Hypothyroid	10
Diabetes Mellitus	8
Short Stature	2
Delayed Puberty / Hypogonadism	2
Total	22

8.5.2 Iron Deposition in Heart and Liver Complications

A total of 110 patients had their liver and cardiac MRI T2* scan completed. Based on Table 8.15, 102 patients (92.73%) had a normal cardiac iron loading and only two patients (1.82%) had severe cardiac iron loading. Only 10 patients (9.09%) had normal liver iron loading. A majority of 43 patients (39.09%) had severe liver iron loading.

Table 8.15: Distribution of Patients in Negeri Sembilan According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Tuanku Ja'afar	97	91	82.73	1	0.91	3	2.73	2	1.82
Hospital Tuanku Ampuan Najihah	12	11	10.00	1	0.91	0	0.00	0	0.00
Hospital Jelebu	1	0	0.00	0	0.00	1	0.91	0	0.00
Total	110	102	92.73	2	1.82	4	3.64	2	1.82

Table 8.16: Distribution of Patients in Negeri Sembilan According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Tuanku Ja'afar	96	10	9.09	16	14.55	34	30.91	36	32.73
Hospital Tuanku Ampuan Najihah	13	0	0.00	2	1.82	5	4.55	6	5.45
Hospital Jelebu	1	0	0.00	0	0.00	0	0.00	1	0.91
Total	110	10	9.09	18	16.36	39	35.45	43	39.09

8.5.3 Deaths Cases

A total of 13 deceased patients had been reported in Negeri Sembilan. The cause of death is shown in Table 8.17 and the commonest cause of death was due to infections. One patient died with upper gastrointestinal bleeding due to portal hypertension secondary to chronic hepatitis C (liver disease).

Table 8.17: Cumulative Known Causes of Death in Negeri Sembilan

Causes of Death	Number of Patients (n)
Infections	8
Cardiac	2
Endocrine Complications	1
Liver Disease	1
Motor Vehicle Accident (MVA)	1
Total	13

8.6 CONCLUSION

In 2020, there is a 15.35% increment as compared to 2019 in the total number of living thalassaemia patients registered in Negeri Sembilan.

Most of the newly diagnosed paediatric patients who needed iron chelation therapy are prescribed DFX monotherapy. Currently, there are 27 patients on combination therapy. TDT patients made up the majority of the ones prescribed with iron-chelation therapy at 102 patients (99.03%). There are only 27 NTDT patients (20.77%) that are prescribed iron chelation therapy.

In Negeri Sembilan, 22 of the thalassaemia patients have endocrine complications. A majority of them were diagnosed with hypothyroid and currently treated with Levothyroxine.

The liver and cardiac MRI T2* scan is only done in Hospital Tuanku Ja'afar. There are only 110 patients who completed their scan. A majority of the patients have normal cardiac iron loading with 102 patients (92.73%) and 43 patients (39.09%) have severe liver iron loading.

Most of the patients with ferritin levels above 5,000 ng/mL are adults who are not compliant to the prescribed iron chelation therapy. Other than that, 53.13% of patients had serum ferritin less than 2,500 ng/mL. As a result, the table shows that there was an improvement of 6.51%.



9.0

PAHANG

9.1 INTRODUCTION

In Pahang, nine government hospitals are included in the registry, namely, Hospital Tengku Ampuan Afzan, Hospital Sultan Haji Ahmad Shah, Hospital Raub, Hospital Kuala Lipis, Hospital Jerantut, Hospital Jengka, Hospital Bentong, Hospital Pekan and Hospital Rompin. According to the Department of Statistics Malaysia, the total population in Pahang is 1.67 million as of 2020.

Transfusion facility has markedly improved in Hospital Tengku Ampuan Afzan and Hospital Sultan Haji Ahmad Shah, where all patients may receive blood transfusion in day care units and receive filtered and nucleic acid test (NAT) blood. Other district hospitals still require ward admissions for transfusion and also receive filtered blood. Hospital Tengku Ampuan Afzan had provided facility for MRI T2* scan facility. Bone densitometry is also provided under the Radiology Department.

9.2 PATIENT DEMOGRAPHICS

Data analysis were taken from patients with status of either alive, lost to follow-up or cured by transplant, and excludes deceased patients. The total number of living patients in Pahang are 458 patients, three deaths were reported in 2020. Two out of three deaths were due to infection and one due to cardiac cause. Regular patients are 233 (50.87%) and irregular patients are 225 (49.13%).

Table 9.1: Distribution of Patients in Pahang by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Tengku Ampuan Afzan	266	58.08
Hospital Sultan Haji Ahmad Shah	133	29.04
Hospital Kuala Lipis	20	4.37
Hospital Pekan	19	4.15
Hospital Bentong	7	1.53
Hospital Raub	6	1.31
Hospital Muadzam Shah	5	1.09
Hospital Jerantut	2	0.44
Total	458	100.00

The majority of patients received their treatment at tertiary hospitals which are Hospital Tengku Ampuan Afzan with 266 patients (58.08%) and Hospital Sultan Haji Ahmad Shah with 133 patients (29.04%).

Table 9.2: Distribution of Patients in Pahang by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	399
Cured by Stem Cell Therapy	3
Total	402
Lost to Follow Up	56
Total	458
Death in 2020	3
Cumulative Reported Deaths	53

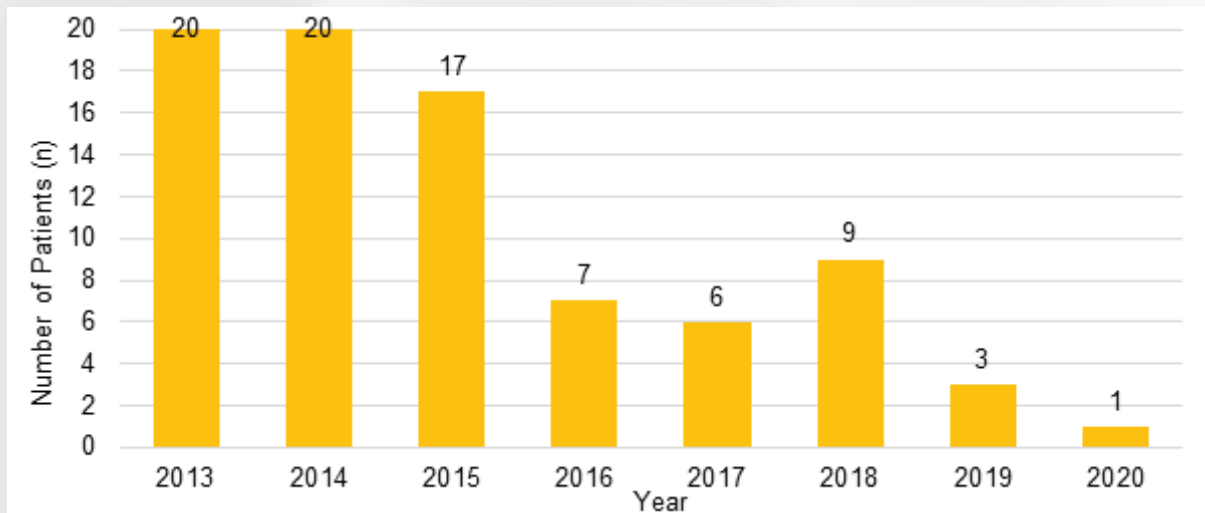


Figure 9.1: Distribution of Thalassaemia Births in Pahang by Year

9.2.1 Age Group

Figures 9.2 indicates that patients aged 35 years and below form 83.62% (383 out of 458 patients) of the thalassaemia cases in Pahang. The eldest patient in Pahang is a 76-years-old diagnosed with HBE/ β -Thalassaemia.

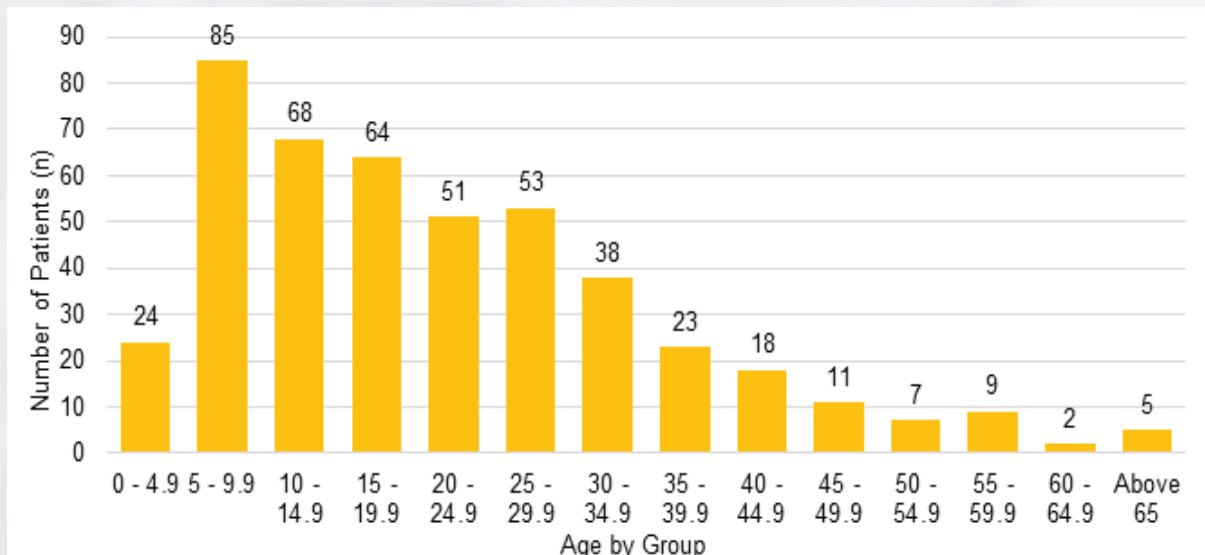


Figure 9.2: Distribution of Patients in Pahang by Age Group

Table 9.3: Distribution of Patients in Pahang According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0-14.9	177	β -Thalassaemia Major	25	14.12
		β -Thalassaemia Intermedia	11	6.21
		HBE/ β -Thalassaemia	68	38.42
		Hb H Disease	56	31.64
		Others	17	9.60
15-29.9	168	β -Thalassaemia Major	31	18.45
		β -Thalassaemia Intermedia	11	6.55
		HBE/ β -Thalassaemia	83	49.40
		Hb H Disease	34	20.24
		Others	9	5.36
30-44.9	79	β -Thalassaemia Major	14	17.72
		β -Thalassaemia Intermedia	6	7.59
		HBE/ β -Thalassaemia	35	44.30
		Hb H Disease	15	18.99
		Others	9	11.39

45-59.9	27	β-Thalassaemia Major	3	11.11
		β-Thalassaemia Intermedia	6	22.22
		HBE/β-Thalassaemia	8	29.63
		Hb H Disease	6	22.22
		Others	4	14.81
60 and above	7	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HBE/β-Thalassaemia	2	28.57
		Hb H Disease	4	57.14
		Others	1	14.29
Total			458	

9.2.2 Gender

Table 9.4 shows that female patients dominate the population of thalassaemia patients in Pahang.

Table 9.4: Distribution of Patients in Pahang According to Gender by Centre

Centre	Total	Male		Female	
		No.	%	No.	%
Hospital Tengku Ampuan Afzan	266	107	23.36	159	34.72
Hospital Sultan Haji Ahmad Shah	133	69	15.07	64	13.97
Hospital Kuala Lipis	20	11	2.40	9	1.97
Hospital Pekan	19	9	1.97	10	2.18
Hospital Bentong	7	3	0.66	4	0.87
Hospital Raub	6	2	0.44	4	0.87
Hospital Muadzam Shah	5	3	0.66	2	0.44
Hospital Jerantut	2	0	0.00	2	0.44
Total	458	204	44.54	254	55.46

9.2.3 Ethnic Group

Based on Figure 9.3, a majority of patients in Pahang are Malays with 413 patients (90.17%), followed by Chinese with 28 patients (6.11%) and other ethnicities with 17 patients (3.71%).

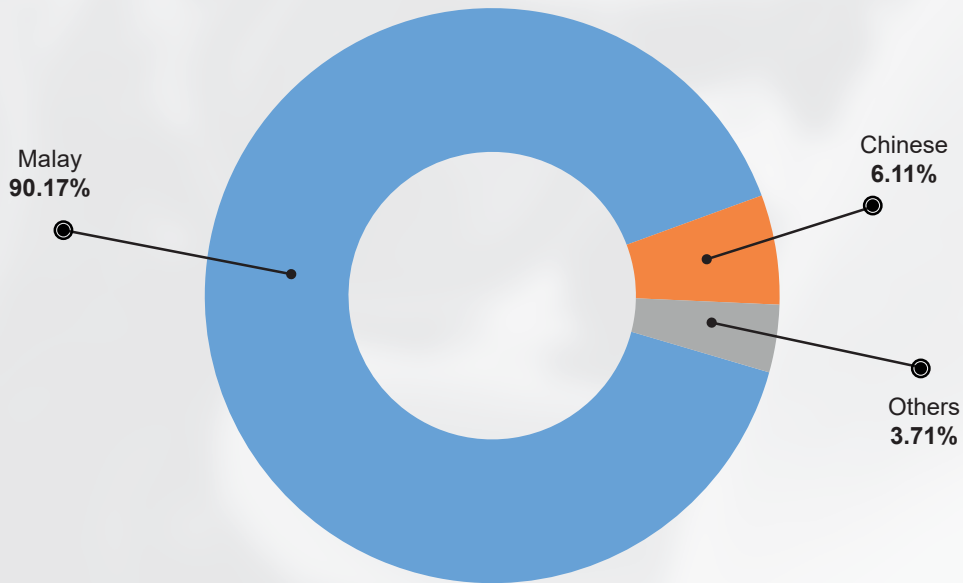


Figure 9.3: Distribution of Patients in Pahang by Ethnic Group

Table 9.5: Distribution of Patients in Pahang According to Ethnic Group by Centre

Centre	Total Number of Patients	Malay		Chinese		Others	
		No.	%	No.	%	No.	%
Hospital Tengku Ampuan Afzan	266	246	53.71	17	3.71	3	0.66
Hospital Sultan Haji Ahmad Shah	133	118	25.76	6	1.31	9	1.97
Hospital Kuala Lipis	20	19	4.15	0	0.00	1	0.22
Hospital Pekan	19	18	3.93	0	0.00	1	0.22
Hospital Bentong	7	3	0.66	3	0.66	1	0.22
Hospital Raub	6	4	0.87	1	0.22	1	0.22
Hospital Muadzam Shah	5	3	0.66	1	0.22	1	0.22
Hospital Jerantut	2	2	0.44	0	0.00	0	0.00
Total	458	413	90.17	28	6.11	17	3.71

9.3 DIAGNOSIS

As shown in Figure 9.4, the diagnosis with the highest number of patients in Pahang is HbE/ β -thalassaemia with 196 patients (42.79%). Meanwhile, the diagnosis with lowest number of patients is β -thalassaemia intermedia with 34 patients (7.42%).

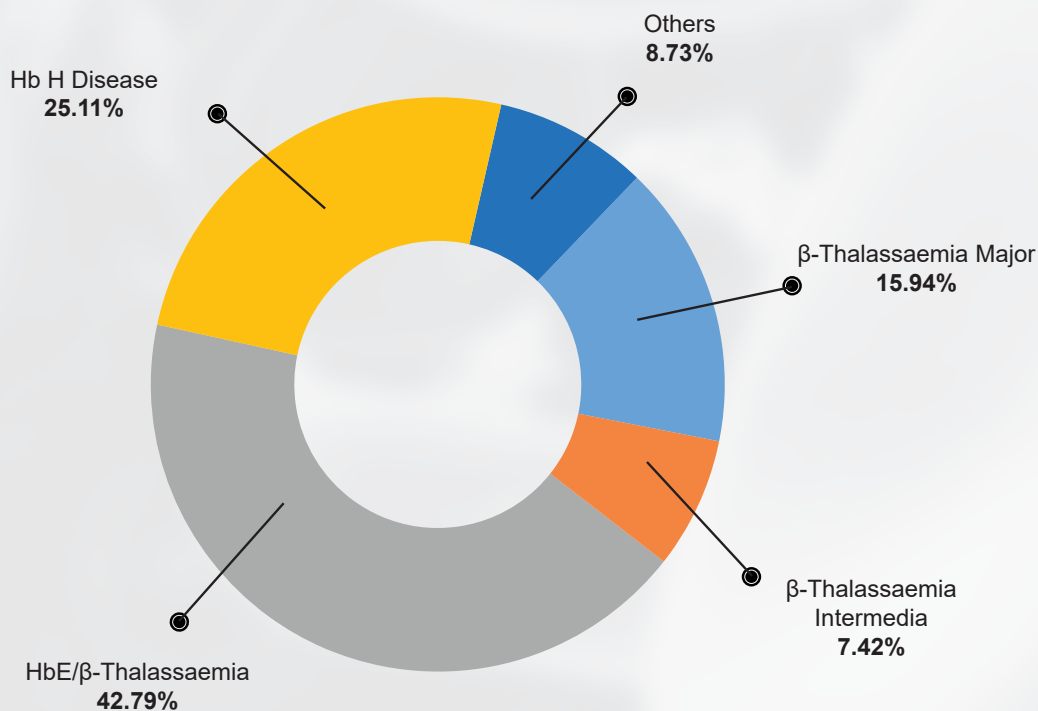


Figure 9.4: Distribution of Patients in Pahang by Diagnosis

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

Centre	Total Number of Patients	β-Thalassaemia major		β-Thalassaemia intermedia		HbE/β-Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Tengku Ampuan Afzan	266	31	6.77	19	4.15	122	26.64	67	14.63	27	5.90
Hospital Sultan Haji Ahmad Shah	133	26	5.68	13	2.84	55	12.01	32	6.99	7	1.53
Hospital Kuala Lipis	20	12	2.62	1	0.22	5	1.09	0	0.00	2	0.44
Hospital Pekan	19	1	0.22	0	0.00	7	1.53	9	1.97	2	0.44
Hospital Bentong	7	1	0.22	0	0.00	2	0.44	4	0.87	0	0.00
Hospital Raub	6	1	0.22	0	0.00	2	0.44	2	0.44	1	0.22
Hospital Muadzam Shah	5	1	0.22	1	0.22	2	0.44	0	0.00	1	0.22
Hospital Jerantut	2	0	0.00	0	0.00	1	0.22	1	0.22	0	0.00
Total	458	73	15.94	34	7.42	196	42.79	115	25.11	40	8.73

Table 9.7: 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	73	Malay	64	13.97
		Chinese	8	1.75
		Others	1	0.22
β-Thalassaemia Intermedia	34	Malay	31	6.77
		Chinese	1	0.22
		Others	2	0.44
HBE/β-Thalassaemia	196	Malay	182	39.74
		Chinese	9	1.97
		Others	5	1.09
Hb H Disease	115	Malay	101	22.05
		Chinese	7	1.53
		Others	7	1.53
Others	40	Malay	35	7.64
		Chinese	3	0.66
		Others	2	0.44
Total			458	100.00

9.4 TREATMENT

9.4.1 Iron Chelation Therapy

Based on Table 9.8, a total of 267 patients were prescribed iron chelator. There were 198 patients (74.15%) prescribed a monotherapy. Meanwhile, 69 patients (25.84%) were prescribed a combination of iron chelator included three patients were prescribed to a combination of all iron chelators.

Table 9.8: Distribution of Patients in Pahang by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	27	10.11
DFP only	115	43.07
DFX only	56	20.97
DFO + DFP	44	16.48
DFP + DFX	8	3.00
DFO + DFX	14	5.24
DFO + DFP + DFX	3	1.12
Total	267	100.00

Table 9.9: 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	153	DFO only	13	4.87
		DFP only	70	26.22
		DFX only	28	10.49
		DFO + DFP	27	10.11
		DFP + DFX	2	0.75
		DFO + DFX	10	3.75
		DFO + DFP + DFX	3	1.12
Hospital Sultan Haji Ahmad Shah	88	DFO only	13	4.87
		DFP only	37	13.86
		DFX only	12	4.49
		DFO + DFP	16	5.99
		DFP + DFX	6	2.25
		DFO + DFX	4	1.50
		DFO + DFP + DFX	0	0.00
Hospital Kuala Lipis	15	DFO only	1	0.37
		DFP only	4	1.50
		DFX only	9	3.37
		DFO + DFP	1	0.37
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Pekan	3	DFO only	0	0.00
		DFP only	2	0.75
		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
Hospital Raub	4	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	4	1.50
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Bentong	3	DFO only	0	0.00
		DFP only	1	0.37
		DFX only	2	0.75
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Rompin	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
Total			267	100.00

Table 9.10: 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	72	DFO only	2	2.78
		DFP only	24	33.33
		DFX only	39	54.17
		DFO + DFP	0	0.00
		DFP + DFX	5	6.94
		DFO + DFX	1	1.39
		DFO + DFP + DFX	1	1.39
15-29.9	118	DFO only	19	16.10
		DFP only	48	40.68
		DFX only	13	11.02
		DFO + DFP	24	20.34
		DFP + DFX	2	1.69
		DFO + DFX	10	8.47
		DFO + DFP + DFX	2	1.69
30-44.9	55	DFO only	5	9.09
		DFP only	27	49.09
		DFX only	3	5.45
		DFO + DFP	16	29.09
		DFP + DFX	1	1.82
		DFO + DFX	3	5.45
		DFO + DFP + DFX	0	0.00
45-59.9	18	DFO only	1	5.56
		DFP only	12	66.67
		DFX only	1	5.56
		DFO + DFP	4	22.22
		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			267	

9.4.2 Serum Ferritin Level

Table 9.11 shows that there are 135 TDT patients who had their serum ferritin level measured in 2020. A total of 88 patients (65.19%) has serum ferritin level below 2,499 ng/mL and 47 patients (34.81%) have a serum ferritin level above 2,500 ng/mL.

Table 9.11: Distribution of TDT 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	73	21	15.56	30	22.22	15	11.11	6	4.44	1	0.74
Hospital Sultan Haji Ahmad Shah	46	11	8.15	12	8.89	14	10.37	5	3.70	4	2.96
Hospital Kuala Lipis	8	1	0.74	5	3.70	1	0.74	0	0.00	1	0.74
Hospital Pekan	4	3	2.22	1	0.74	0	0.00	0	0.00	0	0.00
Hospital Raub	2	0	0.00	2	1.48	0	0.00	0	0.00	0	0.00
Hospital Muadzam Shah	2	1	0.74	1	0.74	0	0.00	0	0.00	0	0.00
Total	135	37	27.41	51	37.78	30	22.22	11	8.15	6	4.44

9.5 COMPLICATIONS AND DEATHS

9.5.1 Complications

Table 9.12 shows 19 patients with endocrine complications. There are nine patients with short stature, followed by eight patients with delayed puberty, one patient had hypothyroid and one patient with hypogonadism.

Table 9.12: Distribution of Patients in Pahang According to Endocrine Complications

Complications	Number of Patients (n)
Short stature	9
Delayed puberty	8
Hypothyroid	1
Hypogonadism	1
Total	19

9.5.2 Iron Deposition in Heart and Liver Complications

A total of 64 patients completed their liver and cardiac MRI T2* scans. Based on Table 9.13, five (7.81%) patients are with severe iron loading, one patient have moderate iron loading and one patient with mild iron loading for cardiac. About 57 patients (89.06%) have normal iron deposition in cardiac.

Based on Table 9.14, a total of 22 patients (34.38%) has severe iron deposition followed by 30 patients (46.88%) who have moderate iron deposition for liver. Only one patient has normal iron deposition.

Table 9.13: Distribution of Patients in Pahang According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Tengku Ampuan Afzan	52	47	73.44	0	0.00	1	1.56	4	6.25
Hospital Sultan Haji Ahmad Shah	12	10	15.63	1	1.56	0	0.00	1	1.56
Total	64	57	89.06	1	1.56	1	1.56	5	7.81

Table 9.14: Distribution of Patients in Pahang According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Tengku Ampuan Afzan	52	1	1.56	9	14.06	27	42.19	15	23.44
Hospital Sultan Haji Ahmad Shah	12	0	0.00	2	3.13	3	4.69	7	10.94
Total	64	1	1.56	11	17.19	30	46.88	22	34.38

9.5.3 Death Cases

According to Table 9.15, there are a cumulative 53 death cases in Pahang including two patients with unknown cause of death. The major cause of death is infection with 28 patients (54.90%) specifically septic shock, multi-organ failure, septic shock with multi-organ failure, and severe sepsis. In 2020, there are 3 death cases were recorded in Hospital Tengku Ampuan Afzan. They were one death case of 89 years old Chinese woman with cardiac and two death cases with infections; a 34 years old man and a 29 years old woman.

Table 9.15: Cumulative Known Causes of Death in Pahang

Causes Of Death	Number of Patients (n)
Infections	28
Cardiac	10
Malignancy	3
Liver disease	2
Bone Marrow Transplant Complications	2
Others	2
Motor Vehicle Accident (MVA)	1
Renal Complications	1
Surgical Complications	1
Died at Home/Brought in Dead to Hospital	1
Total	51

9.6 CONCLUSION

There are 233 TDT patients (50.87%) and 225 NTDT patients (49.13%) in Pahang. In 2020, three patient deaths have been recorded due to infections and cardiac. Patients in the age group of 6-10 years old form the largest number of patients. The number of female patients is higher than male patients in Pahang. Based on ethnicity, Malays had the highest number of 413 patients and the majority of them were diagnosed with HbE/ β -Thalassaemia and Hb H Disease. This should be explained with the majority of population in Pahang is Malay.

Patients that are diagnosed with HbE/ β -Thalassaemia are the largest group with 196 patients (42.79%), Hb H Disease had 115 patients (25.11%), β -Thalassaemia Major had 73 patients (15.94%), other diagnosis with 40 patients (8.73%) and β -Thalassaemia Intermedia had 34 patients (7.42%). There are 267 patients who received iron chelating agents and only three patients were prescribed a combination of three iron chelators. Based on 135 patients who have their serum ferritin level measured in 2020, 88 patients (65.19%) have a serum ferritin level below 2,500 ng/mL and 47 patients (34.81%) have a serum ferritin above 2,500 ng/mL.

The most common endocrine complications are short stature with nine patients and delayed puberty with eight patients. For liver and cardiac MRI T2* scan, 64 patients have the results. There are five patients with severe and 57 patients with normal iron loading in cardiac. For the liver, 22 patients are with severe and only one patient with normal iron loading in liver.



10.0
PERAK

10.1 INTRODUCTION

Perak is the fourth largest state in the country and the second largest state in Peninsular Malaysia. It has an estimated population of 2.5 million people (Department of Statistics Malaysia, 2019).

In Perak, thalassaemia patients are only treated in Hospital Raja Permaisuri Bainun (Ipoh), Hospital Taiping, Hospital Teluk Intan, Hospital Manjung, Hospital Slim River, Hospital Kuala Kangsar and Hospital Gerik. Hospital Ipoh and Hospital Taiping carry out transfusions in their Ambulatory Care Centre (ACC). For hospitals without Ambulatory Care Centre, the transfusion is carried out in the wards.

10.2 PATIENT DEMOGRAPHIC

There are 602 thalassaemia patients in Perak. These patients receive transfusions and care in the seven hospitals listed in Table 10.1. The data analysed were taken from patients who are either alive, lost to follow-up or cured by transplant. Table 10.2 shows a total of 552 living patients, 50 lost to follow-up and 51 cumulative reported deaths.

Table 10.1: Distribution of Patients in Perak by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Raja Permaisuri Bainun	359	59.63
Hospital Taiping	102	16.94
Hospital Teluk Intan	56	9.30
Hospital Seri Manjung	45	7.48
Hospital Kuala Kangsar	14	2.33
Hospital Slim River	13	2.16
Hospital Gerik	13	2.16
Total	602	100.00

Table 10.2: Distribution of Patients in Perak by Vital Status

Vital Status	Number of Patients (n)
Alive and on Active Treatment	544
Cured by Stem Cell Therapy	8
Total	552
Lost to Follow Up	50
Total	602
Deaths in 2020	4
Cumulative Reported Deaths	51

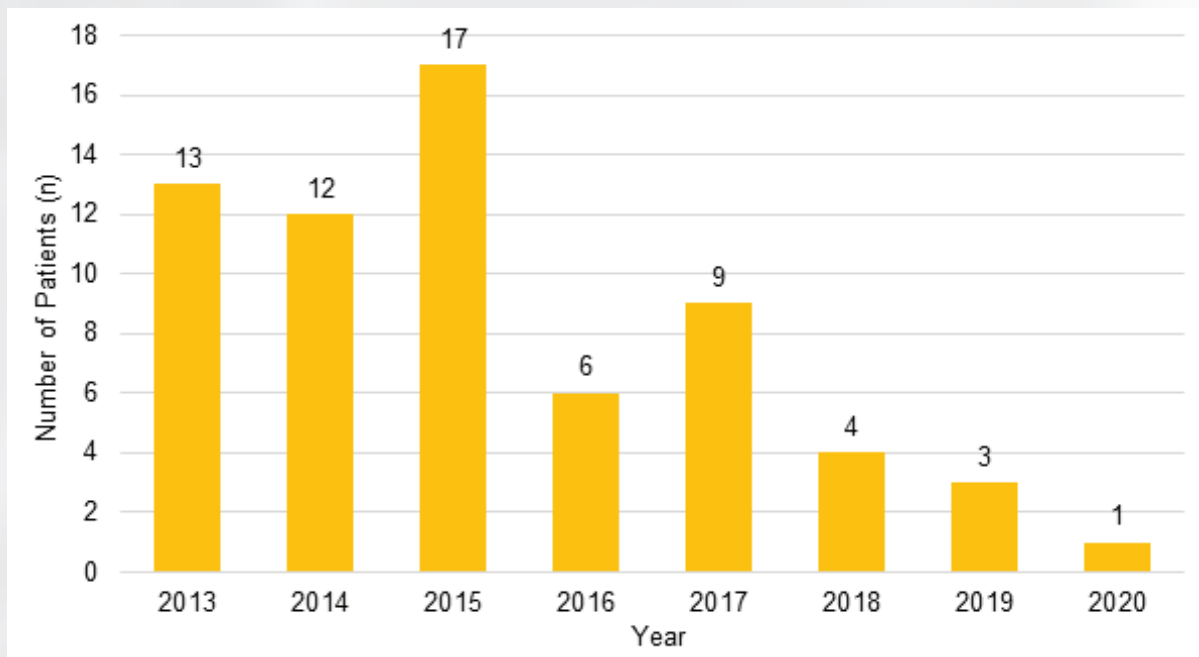


Figure 10.1: Distribution of Thalassaemia Births in Perak by Year

10.2.1 Age Groups

Based on Figure 10.2, 42.19% of thalassaemia patients in Perak are 20 years old and below, and are in the paediatric adolescent age groups. Patients above 20 years old account for 57.81% of the thalassaemic in Perak. There is a drop in total number of patients in the age group 15-19.9 likely due to missing data.

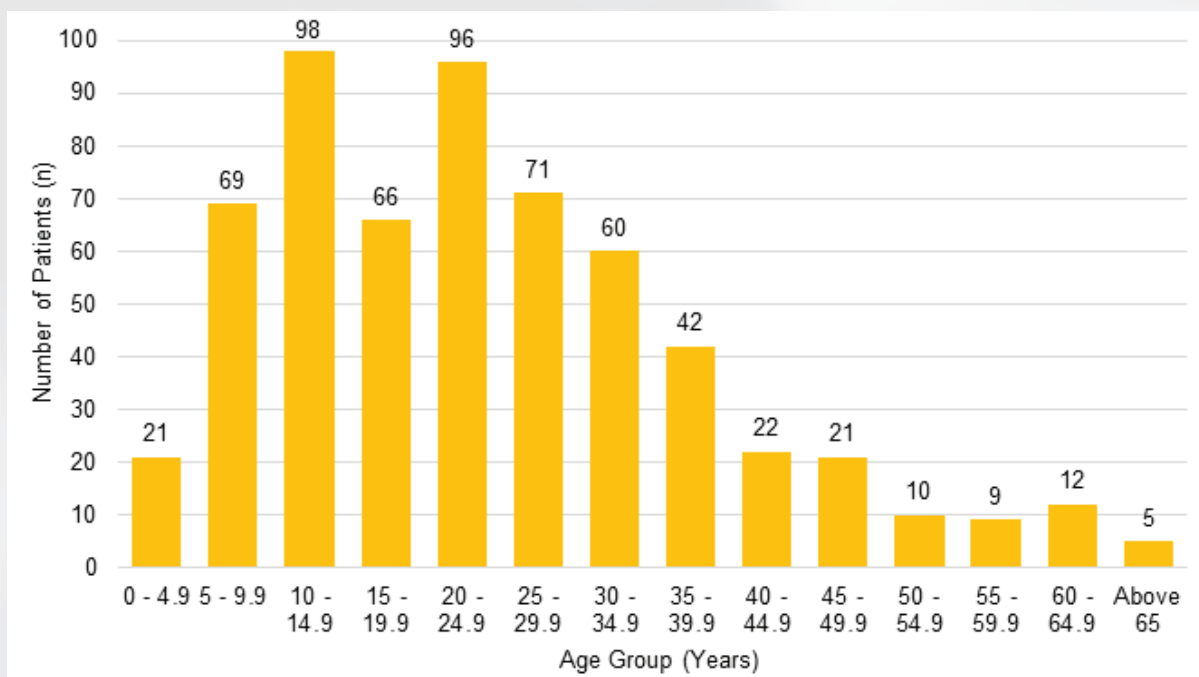


Figure 10.2: Distribution of Patients in Perak by Age Group

Table 10.3: Distribution of Patients in Perak According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0-14.9	188	β-Thalassaemia Major	35	18.62
		β-Thalassaemia Intermedia	20	10.64
		HbE β-Thalassaemia	93	49.47
		Hb H Disease	36	19.15
		Others	4	2.13
15-29.9	233	β-Thalassaemia Major	55	23.61
		β-Thalassaemia Intermedia	20	8.58
		HbE β-Thalassaemia	109	46.78
		Hb H Disease	45	19.31
		Others	4	1.72
30-44.9	124	β-Thalassaemia Major	22	17.74
		β-Thalassaemia Intermedia	10	8.06
		HbE β-Thalassaemia	49	39.52
		Hb H Disease	37	29.84
		Others	6	4.84
45-59.9	40	β-Thalassaemia Major	1	2.50
		β-Thalassaemia Intermedia	3	7.50
		HbE β-Thalassaemia	17	42.50
		Hb H Disease	17	42.50
		Others	2	5.00
60 and above	17	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	2	11.76
		HbE β-Thalassaemia	4	23.53
		Hb H Disease	10	58.82
		Others	1	5.88
Total			602	

10.2.2 Gender

Table 10.4 shows the distribution of thalassaemia patients in Perak by gender. There are 313 male patients (51.99%) and 289 female patients (48.01%).

Table 10.4: Distribution of Patients in Perak According to Gender by Centre

Centre	Total Number of Patients	Male		Female	
		No.	%	No.	%
Hospital Raja Permaisuri Bainun	359	172	28.57	187	31.06
Hospital Taiping	102	56	9.30	46	7.64
Hospital Teluk Intan	56	38	6.31	18	2.99
Hospital Seri Manjung	45	25	4.15	20	3.32
Hospital Kuala Kangsar	14	7	1.16	7	1.16
Hospital Slim River	13	8	1.33	5	0.83
Hospital Gerik	13	7	1.16	6	1.00
Total	602	313	51.99	289	48.01

10.2.3 Ethnic Group

Malay patients covered the largest cohort of thalassaemics with 455 patients (75.58%), followed by the Chinese (117 patients, 19.44%). Other ethnic groups contributed to a minor proportion of the total number of thalassaemic in Perak and this includes Indian (three patients, 0.50%), Orang Asli (22 patients, 3.65%) and other ethnic groups (five patients, 0.83%).

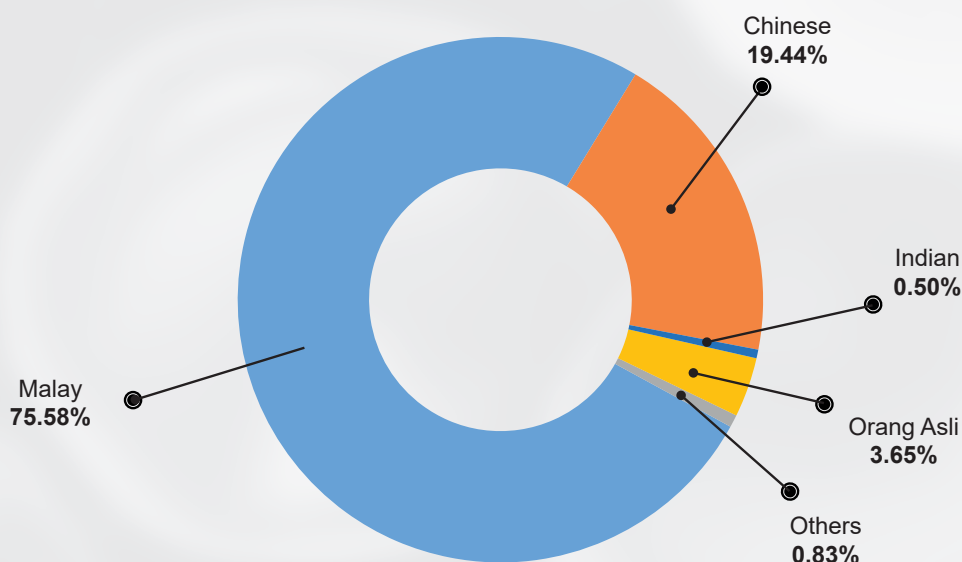


Figure 10.3: Distribution of Patients in Perak by Ethnic Group

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

Centre	Total Number of Patients	Malay		Chinese		Indian		Orang Asli		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Raja Permaisuri Bainun	359	242	40.20	99	16.45	2	0.33	15	2.49	1	0.17
Hospital Taiping	102	94	15.61	8	1.33	0	0.00	0	0.00	0	0.00
Hospital Teluk Intan	56	44	7.31	6	1.00	1	0.17	4	0.66	1	0.17
Hospital Seri Manjung	45	40	6.64	4	0.66	0	0.00	0	0.00	1	0.17
Hospital Kuala Kangsar	14	14	2.33	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Slim River	13	11	1.83	0	0.00	0	0.00	0	0.00	2	0.33
Hospital Gerik	13	10	1.66	0	0.00	0	0.00	3	0.50	0	0.00
Total	602	455	75.58	117	19.44	3	0.50	22	3.65	5	0.83

10.3 DIAGNOSIS

HbE/ β -thalassaemia is the commonest diagnosis in Perak with 272 patients (45.18%), followed by Hb H disease (145 patients, 24.09%), β -thalassaemia major (113 patients, 18.77%) and β -thalassaemia intermedia (55 patients, 9.14%). The remaining 17 patients (2.82%) have other forms of haemoglobinopathy.

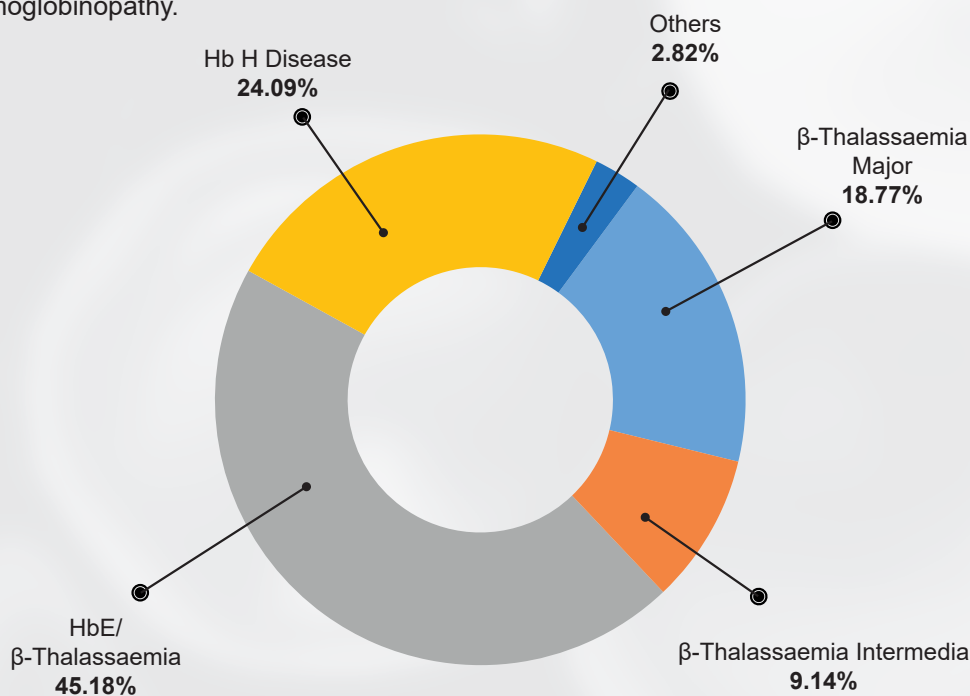


Figure 10.4: Distribution of Patients in Perak by Diagnosis

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

Centre	Total Number of Patients	β-Thalassaemia major		β-Thalassaemia intermedia		HbE/β-Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Raja Permaisuri Bainun	359	52	8.64	35	5.81	146	24.25	114	18.94	12	1.99
Hospital Taiping	102	22	3.65	9	1.50	55	9.14	13	2.16	3	0.50
Hospital Teluk Intan	56	8	1.33	8	1.33	33	5.48	6	1.00	1	0.17
Hospital Seri Manjung	45	20	3.32	2	0.33	15	2.49	7	1.16	1	0.17
Hospital Kuala Kangsar	14	4	0.66	0	0.00	10	1.66	0	0.00	0	0.00
Hospital Slim River	13	5	0.83	1	0.17	7	1.16	0	0.00	0	0.00
Hospital Gerik	13	2	0.33	0	0.00	6	1.00	5	0.83	0	0.00
Total	602	113	18.77	55	9.14	272	45.18	145	24.09	17	2.82

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	197	Malay	73	12.13
		Chinese	38	6.31
		Indian	1	0.17
		Orang Asli	0	0.00
		Others	1	0.17
β-Thalassaemia Intermedia	41	Malay	43	7.14
		Chinese	10	1.66
		Indian	1	0.17
		Orang Asli	1	0.17
		Others	0	0.00
HbE/β-Thalassaemia	300	Malay	235	39.04
		Chinese	21	3.49
		Indian	0	0.00
		Orang Asli	12	1.99
		Others	4	0.66
Hb H Disease	122	Malay	94	15.61
		Chinese	44	7.31
		Indian	0	0.00
		Orang Asli	7	1.16
		Others	0	0.00
Others	17	Malay	10	1.66
		Chinese	4	0.66
		Indian	1	0.17
		Orang Asli	2	0.33
		Others	0	0.00
Total			602	100.00

10.4 TREATMENT

10.4.1 Iron Chelation Therapy

Based on Table 10.8, 354 patients in Perak received iron chelation therapy. A total of 84 patients (23.73%) were prescribed DFO, and 79 patients (22.32%) were prescribed oral DFP monotherapy. Additionally, 129 patients (36.44%) are on DFX monotherapy, which is normally reserved for patients who could not tolerate other chelating agents. Meanwhile, 47 patients (13.28%) are receiving a combination of DFO and DFP. The number of patients with a combination iron chelator are six patients (1.69%) who were prescribed DFP+DFX and eight patients (2.26%) who were prescribed DFO+DFX. Only one patient (0.28%) is prescribed a combination of three chelators.

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

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	84	23.73
DFP only	79	22.32
DFX only	129	36.44
DFO + DFP	47	13.28
DFP + DFX	6	1.69
DFO + DFX	8	2.26
DFO + DFP + DFX	1	0.28
Total	354	100.00

Table 10.9: Distribution of Patients in Perak by The Type of Transfusion Status

Transfusion Status	Total Number of Patients	With Iron Chelation Therapy	Percentage (%)	Without Iron Chelation Therapy	Percentage (%)
TDT	296	271	91.55	25	8.45
NTDT	306	83	27.12	223	72.88
Total	602	354	58.80	248	41.20

Table 10.10: 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	43	12.15
		DFP only	52	14.69
		DFX only	54	15.25
		DFO + DFP	24	6.78
		DFP + DFX	3	0.85
		DFO + DFX	3	0.85
		DFO + DFP + DFX	1	0.28
Hospital Taiping	70	DFO only	11	3.11
		DFP only	16	4.52
		DFX only	25	7.06
		DFO + DFP	13	3.67
		DFP + DFX	2	0.56
		DFO + DFX	3	0.85
		DFO + DFP + DFX	0	0.00
Hospital Teluk Intan	39	DFO only	11	3.11
		DFP only	5	1.41
		DFX only	18	5.08
		DFO + DFP	5	1.41
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		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	1	0.28
		DFP only	0	0.00
		DFX only	4	1.13
		DFO + DFP	0	0.00

		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Seri Manjung	38	DFO only	11	3.11
		DFP only	4	1.13
		DFX only	16	4.52
		DFO + DFP	4	1.13
		DFP + DFX	1	0.28
		DFO + DFX	2	0.56
		DFO + DFP + DFX	0	0.00
		Total		

Table 10.11: 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	92	DFO only	16	17.39
		DFP only	3	3.26
		DFX only	72	78.26
		DFO + DFP	1	1.09
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15-29.9	155	DFO only	37	23.87
		DFP only	31	20.00
		DFX only	48	30.97
		DFO + DFP	30	19.35
		DFP + DFX	4	2.58
		DFO + DFX	4	2.58
		DFO + DFP + DFX	1	0.65
30-44.9	79	DFO only	26	32.91
		DFP only	28	35.44
		DFX only	7	8.86
		DFO + DFP	13	16.46
		DFP + DFX	1	1.27
		DFO + DFX	4	5.06
		DFO + DFP + DFX	0	0.00
45-59.9	14	DFO only	2	14.29
		DFP only	9	64.29
		DFX only	2	14.29
		DFO + DFP	1	7.14
		DFP + DFX	0	0.00

		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 and above	14	DFO only	3	21.43
		DFP only	8	57.14
		DFX only	0	0.00
		DFO + DFP	2	14.29
		DFP + DFX	1	7.14
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
		Total		

10.4.2 Serum Ferritin Level

Out of 296 TDT patients in 2020, only 175 (59.12%) had their most recent ferritin level measured. A total of 77 patients (44.00%) has serum ferritin level below 2,499 ng/mL. 98 patients (56.00%) have a serum ferritin level above 2,500 ng/mL.

Table 10.12: Distribution of TDT 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	120	19	10.86	35	20.00	47	26.86	19	10.86	0	0.00
Hospital Taiping	31	3	1.71	6	3.43	14	8.00	6	3.43	2	1.14
Hospital Seri Manjung	16	1	0.57	8	4.57	5	2.86	2	1.14	0	0.00
Hospital Slim River	7	5	2.86	0	0.00	2	1.14	0	0.00	0	0.00
Hospital Teluk Intan	1	0	0.00	0	0.00	1	0.57	0	0.00	0	0.00
Hospital Kuala Kangsar	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Gerik	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	175	28	16.00	49	28.00	69	39.43	27	15.43	2	1.14

10.5 COMPLICATIONS AND DEATHS

10.5.1 Complications

The prevalence of severe complications due to iron overload is still high. One of the main reasons for infection in thalassaemia is transfusion-transmitted infection. A wide variety of organisms including bacteria and viruses can be transmitted through blood transfusions. Table 10.13 shows the number of blood-borne transfusion transmissible infections in Perak.

Table 10.13: Distribution of Patients in Perak According to Transfusion Transmissible Infections

Complications	2007-2019	2020
Hepatitis C: Anti HCV	17	0
HCV RNA	0	0
Hepatitis B	0	0
HIV	0	0
Total	17	0

Endocrine complications are also common among thalassaemia patients. Despite good chelation therapy begun early in life, problems such as delayed puberty and short stature may occur. Table 10.14 shows the statistic of endocrine complications among the patients in Perak. There are 31 patients who showed several forms of complications with the highest number being short stature with 18 patients, followed by eight patients with delayed puberty, hypothyroid with three patients and diabetes mellitus with two patients.

Table 10.14: Distribution of Patients in Perak According to Endocrine Complications

Endocrine	Number of Patients (n)
Short Stature	18
Delayed Puberty	8
Hypothyroid	3
Diabetes Mellitus	2
Total	31

10.5.2 Iron Deposition in Heart and Liver Complications

The liver and cardiac complications due to iron overload is a major cause of mortality and morbidity in TDT patients. Patients with high serum ferritin due to blood transfusion have a higher risk of developing complication which can lead to death.

From Table 10.15, 62 patients had their cardiac MRI T2* scan done in 2020. There are 54 patients (87.10%) with normal grade, one patient (1.61%) with mild grade, two patients (3.23%) with moderate grade and five patients (8.06%) presented with severe grade of iron loading in the cardiac.

Table 10.15: Distribution of Patients in Perak According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Raja Permaisuri Bainun	62	54	87.1	1	1.61	2	3.23	5	8.06
Total	62	54	87.1	1	1.61	2	3.23	5	8.06

Based on Table 10.16, the liver MRI results showed that only 10 patients (16.13 %) had normal iron content, 28 patients (45.16%) with mild iron content, 20 patients (32.26%) had moderate iron content and four patients (6.45%) had severe iron content in their liver.

Table 10.16: Distribution of Patients in Perak According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Raja Permaisuri Bainun	62	10	16.13	28	45.16	20	32.26	4	6.45
Total	62	10	16.13	28	45.16	20	32.26	4	6.45

10.5.3 Deaths Cases

Based on Table 10.17, the total cumulative cause of death in Perak is 51 patients included two cases with unknown cause of death. Cardiac cause leads the overall causes with 26 patients, followed by infection with 14 patients. In 2020, there were four deaths and the causes were due to cardiac specifically chronic iron overload (two cases) and infection specifically septic shock with two cases.

Table 10.17: Cumulative Known Causes of Death in Perak

Causes of Death	Number of Patients (n)
Cardiac	26
Infections	14
Died at Home/Brought in Dead to Hospital	5
Endocrine Complications	2
Malignancy	1
Others	1
Total	49

10.6 CONCLUSION

This chapter provides a brief overview on the number of thalassaemia patients, who received treatment in the state of Perak. In 2020, the number of thalassaemia patients reported was 602, of which TDT and NTDT comprised 296 (49.17%) and 306 (50.83%) patients, respectively.

The majority of thalassaemia patients in Perak are adults (348 patients, 57.81%) followed by paediatric (254 patients, 42.19%). Malay patients constitute 75.58% of the total number of patients, followed by Chinese (19.44%), and other ethnicities (4.98%). The subtypes of thalassaemia found in the state are mainly HbE/ β -thalassaemia (45.18%), followed by Hb H disease (24.09%), β -thalassaemia major (18.77%), β -thalassaemia intermedia (9.14%) and other subtypes (2.8%).

The total number of patients prescribed with iron chelation therapy is 354 (58.80%), which includes TDT and NTDT patients. A total of 271 TDT patients (91.55%) are currently receiving chelation therapy. The remaining 25 (8.45%) of TDT patients are not on chelation therapy, mainly due to non-compliance to treatment. A total of 83 NTDT patients (27.12%) are also receiving chelation therapy. The main type of chelator used is DFX (36.44%), followed by DFO (23.73%). 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 (13.28%).

Serum ferritin levels below 1,000 ng/mL indicate good compliance to iron chelators, however, only 16.0% of TDT patients fall into this category. A total of 175 TDT patients (59.12%) had their serum ferritin levels measured, of which 44.00% have an acceptable serum ferritin level of lower than 2,500 ng/mL. Thus, compliance still remains an issue in 98 patients (56.00%) 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.



11.0
PERLIS

11.1 INTRODUCTION

Perlis is the smallest state in Malaysia with an estimated population of 254,900 (Department of Statistics Malaysia, 2020). There is only one hospital in Perlis, which is Hospital Tunku Fauziah located in Kangar.

11.2 PATIENT DEMOGRAPHICS

Data analysed were taken from patients who are either alive, lost to follow up, cured by stem cell therapy and deceased patients. There are 134 living patients in Perlis, and 14 patients are deceased. There is no death reported among thalassaemia patients in 2020.

Table 11.1: Distribution of Patients in Perlis by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Tunku Fauziah	134	100.00
Total	134	100.00

Table 11.2: Distribution of Patients in Perlis by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	118
Cured by Stem Cell Therapy	1
Total	119
Lost to Follow Up	15
Total	134
Death in 2020	0
Cumulative Reported Death	14

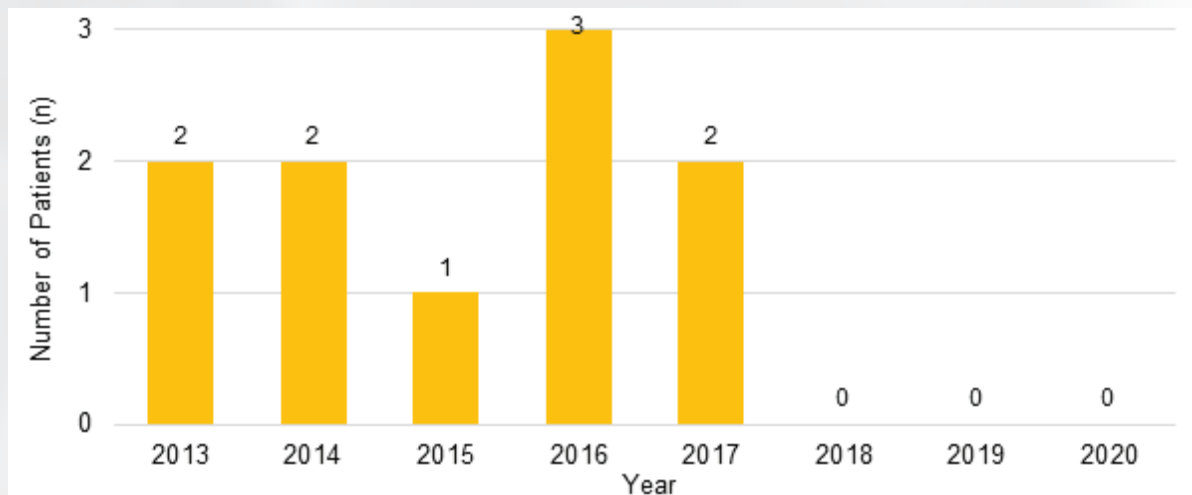


Figure 11.1: Distribution of Thalassaemia Births in Perlis by Year

11.2.1 Age Group

Based on Figure 11.2, the youngest patient in Perlis is three years old and the oldest patient is 65 years old. Sixty-five patients (48.50%) are below 20 years old.

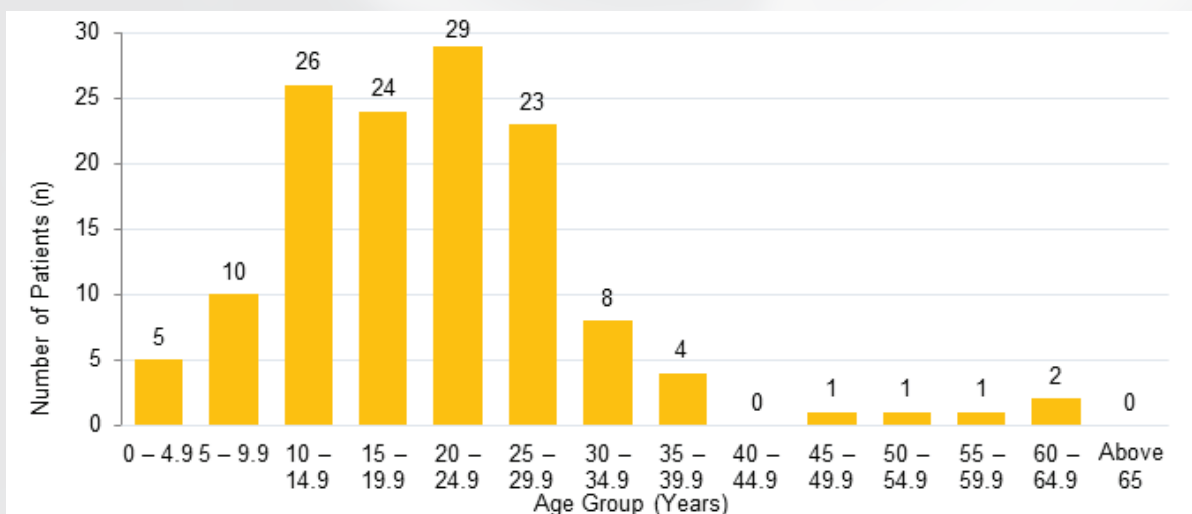


Figure 11.2: Distribution of Patients in Perlis by Age Group

Table 11.3: Distribution of Patients in Perlis According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0–14.9	41	β- Thalassaemia Major	8	19.51
		β-Thalassaemia Intermedia	10	24.39
		HbE/β-Thalassaemia	10	24.39
		Hb H Disease	11	26.83
		Others	2	4.88
15-29.9	76	β- Thalassaemia Major	15	19.74
		β-Thalassaemia Intermedia	7	9.21
		HbE/β-Thalassaemia	35	46.05
		Hb H Disease	18	23.68
		Others	1	1.32
30–44.9	12	β- Thalassaemia Major	5	41.67
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	6	50.00
		Hb H Disease	1	8.33
		Others	0	0.00
45–59.9	3	β- Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	2	66.67
		Hb H Disease	0	0.00
		Others	1	33.33
60 and above	2	β- Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	50.00
		Hb H Disease	0	0.00
		Others	1	50.00
Total			134	

11.2.2 Gender

Table 11.4 shows that the total number of patients for female is higher than male.

Table 11.4: Distribution of Patients in Perlis According to Gender

Centre	Total	Male		Female	
		No.	%	No.	%
Hospital Tunku Fauziah	134	66	49.25	68	50.75
Total	134	66	49.25	68	50.75

11.2.3 Ethnic Group

As shown in Figure 11.3, most thalassaemia patients in Perlis are of Malay descent (129 patients, 96.27%). Three patients (2.24%) are Chinese and two patients are Thai (1.49%).

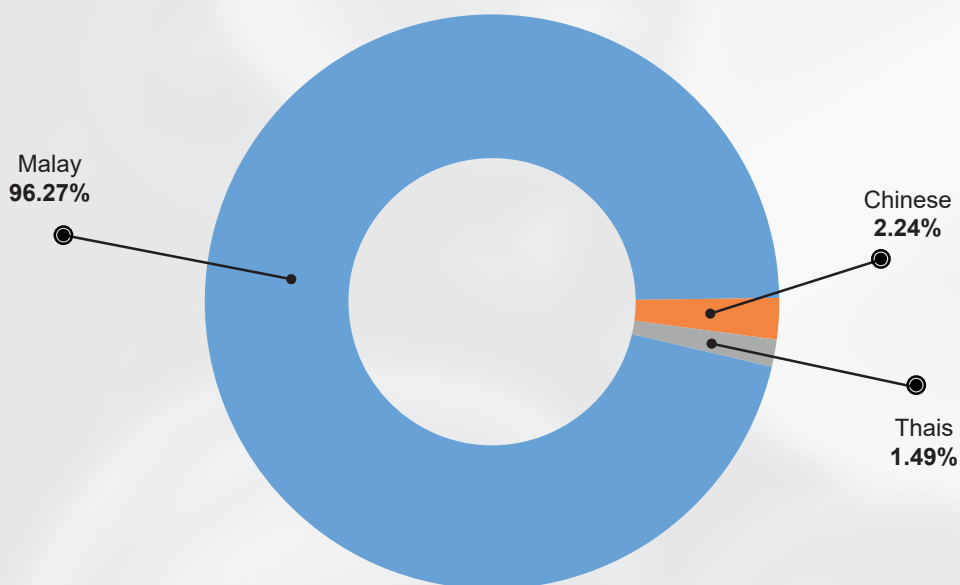


Figure 11.3: Distribution of Patients in Perlis by Ethnic Group

11.3 DIAGNOSIS

Based on Figure 11.4, the commonest diagnosis among patients in Perlis is HbE/ β -thalassaemia with 54 patients (40.30%) followed by Hb H disease with 30 patients (22.39%), β -thalassaemia major with 28 patients (20.90%) and β -thalassaemia intermedia with 17 patients (12.69%). The remaining five patients (3.73%) have other diagnoses.

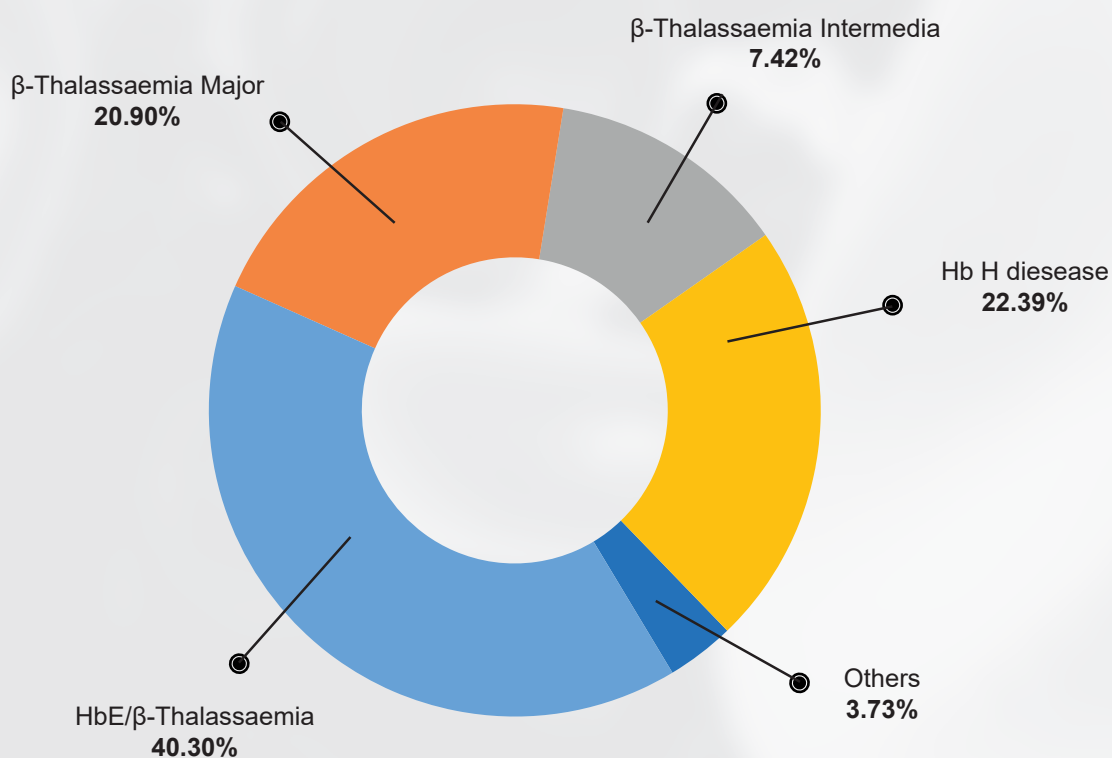


Figure 11.4: Distribution of Patients in Perlis by Diagnosis

Table 11.5: 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	28	Malay	27	20.15
		Chinese	1	0.75
		Thais	0	0.00
		Others	0	0.00
β- Thalassaemia Intermedia	17	Malay	17	12.69
		Chinese	0	0.00
		Thais	0	0.00
		Others	0	0.00
HbE/β- Thalassaemia	54	Malay	52	38.81
		Chinese	1	0.75
		Thais	1	0.75
		Others	0	0.00
Hb H Disease	30	Malay	28	20.90
		Chinese	1	0.75
		Thais	1	0.75
		Others	0	0.00
Others	5	Malay	5	3.73
		Chinese	0	0.00
		Thais	0	0.00
		Others	0	0.00
Total			134	100.00

11.4 TREATMENT

11.4.1 Iron Chelation Therapy

There are 81 patients (60.44%) in Perlis receiving iron chelation therapy. Sixty-one patients (75.31%) are prescribed with oral DFX, 3 patients (3.70%) are prescribed with DFO, 11 patients (13.58%) are prescribed with oral DFP and six patients (7.40%) are prescribed a combination therapy.

Table 11.6: Distribution of Patients in Perlis by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	3	3.70
DFP only	11	13.58
DFX only	61	75.31
DFO + DFP	2	2.47
DFP + DFX	3	3.70
DFO + DFX	1	1.23
DFO + DFP + DFX	0	0.00
Total	81	100.00

Table 11.7: 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 (%)
0-14.9	20	DFO only	0	0.00
		DFP only	2	10.00
		DFX only	17	85.00
		DFO + DFP	1	5.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15-29.9	50	DFO only	3	6.00
		DFP only	7	14.00
		DFX only	37	74.00
		DFO + DFP	2	4.00
		DFP + DFX	1	2.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30-44.9	9	DFO only	0	0.00
		DFP only	1	11.11
		DFX only	5	55.56
		DFO + DFP	0	0.00
		DFP + DFX	2	22.22
		DFO + DFX	1	11.11
		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			81	

11.4.2 Serum Ferritin Level

There are 22 regularly transfused patients in Perlis who had their serum ferritin level measured. Ten patients (45.45%) have serum ferritin level lower than 2,499 ng/mL, and 12 patients (54.45%) with serum ferritin level above 2,500 ng/mL.

Table 11.8: Distribution of TDT 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	22	5	22.73	5	22.73	7	31.82	5	22.73	0	0.00
Total	22	5	22.73	5	22.73	7	31.82	5	22.73	0	0.00

11.5 COMPLICATIONS AND DEATHS

11.5.1 Complications

As of November 2020, there is no transfusion transmitted viral infection reported among thalassaemia patients in Perlis. Based on Table 11.9, 27 out of 71 TDT patients above 15 years old (38.03%) have delayed puberty and 14 patients (19.72%) have short stature.

Table 11.9: Distribution of Patients in Perlis According to Endocrine Complications

Complications	Number of Patients (n)
Delayed Puberty	27
Short Stature	14
Total	41

11.5.2 Iron Deposition in Heart and Liver Complications

As shown in the following Tables 11.10 and 11.11, 73 patients completed their liver and cardiac MRI T2* scan in 2020. Table 11.10 showed that 58 patients (79.45%) with no iron loading, eight patients (10.96%) have mild iron loading, four patients (5.48%) with moderate iron loading, three patients (4.11%) presented severe iron loading in the cardiac.

In Table 11.11, only six patients (8.22%) had normal iron loading, 10 patients (13.70%) with mild iron loading, 16 patients (21.92%) had moderate iron loading and 41 patients (56.16%) with severe iron loading in their liver.

Table 11.10: Distribution of Patients in Perlis According to Cardiac MRI T2*

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Tunku Fauziah	73	58	79.45	8	10.96	4	5.48	3	4.11
Total	73	58	79.45	8	10.96	4	5.48	3	4.11

Table 11.11: Distribution of Patients in Perlis According to Liver MRI

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Tunku Fauziah	73	6	8.22	10	13.70	16	21.92	41	56.16
Total	73	6	8.22	10	13.70	16	21.92	41	56.16

11.5.3 Death Cases

There were 14 death cases reported in Perlis. The most common cause of death was cardiac complication. There was no death reported among thalassaemia patients in 2020.

Table 11.12: Cumulative Known Causes of Death in Perlis

Causes Of Death	Number of Patients (n)
Cardiac	7
Motor Vehicle Accident (MVA)	3
Infections	3
Malignancy	1
Total	14

11.6 CONCLUSION

According to the report, 48.50% patients in Perlis are below 20 years old. The median age group is 20-24.9 years old. Most patients are Malay (96.27%), followed by Chinese (2.24%) and Thais (1.49%). The most common type of thalassaemia in Perlis is HbE/ β -thalassaemia (40.30%), followed by Hb H disease (22.39%), β -thalassaemia major (20.90%) and β -thalassaemia intermedia (12.69%). The majority of patients are prescribed with oral DFX monotherapy with 61 patients (75.31%). Only 22 patients had their serum ferritin reported and majority are the regularly transfused patients. Of these, 45.45% achieved serum ferritin < 2,500 ng/mL.

Out of 71 TDT patients that are above 15 years old, approximately 57.75% have delayed puberty and short stature. The liver and cardiac MRI T2* scans were performed on all chelated patients above 10 years old. Based on cardiac MRI T2* scan, 20.55% of patients have mild to severe cardiac iron loading. For liver MRI scan, there are 10 patients with mild (13.70%), 16 patients with moderate (21.92%), and 41 patients with severe (56.16%) iron loading in their hepatic. The liver and cardiac T2* MRI service currently provided in Hospital Tunku Fauziah and is a great help in the assessment of iron overload. There were no deaths reported in 2020.

12.0

PULAU PINANG

12.1 INTRODUCTION

Pulau Pinang has a population of 1,774,800 in 2020 (Department of Statistics Malaysia). Hospital Pulau Pinang and Hospital Seberang Jaya are the hospitals providing treatment for thalassaemia patients in the region.

12.2 PATIENT DEMOGRAPHICS

Data analysed were taken from patients who were either alive, lost to follow-up or cured by transplant and excludes the deceased patients. Pulau Pinang has a total of 523 alive patients with thalassaemia. Six patients were cured by transplant and 32 patients were lost to follow-up.

Table 12.1: Distribution of Patients in Pulau Pinang by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Pulau Pinang	282	53.92
Hospital Seberang Jaya	241	46.08
Total	523	100.00

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

Vital Status	Number of Patients (n)
Alive and On Active Treatment	485
Cured by Stem Cell Therapy	6
Total	491
Lost to Follow Up	32
Total	523
Death in 2020	2
Cumulative Reported Deaths	18

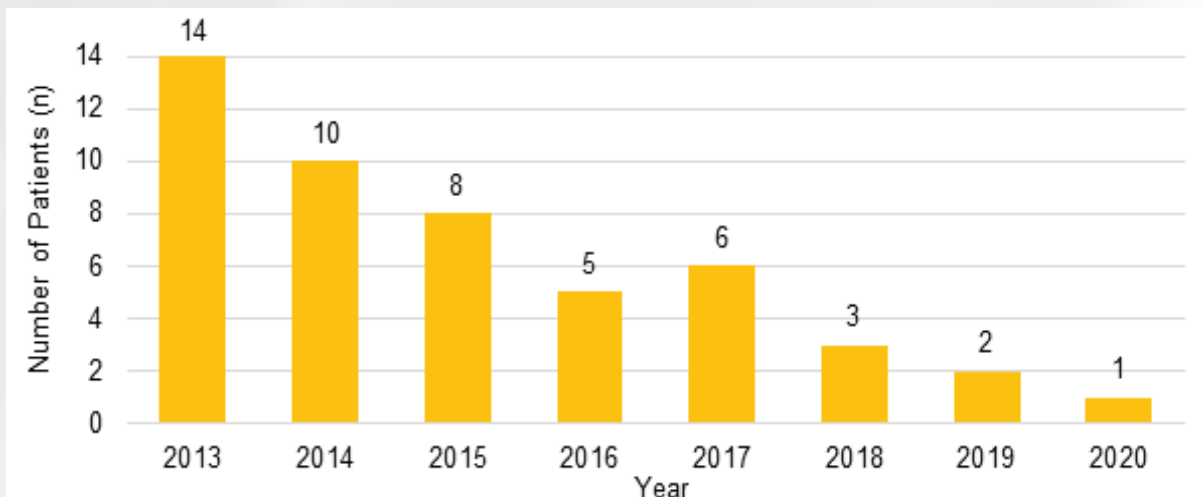


Figure 12.1: Distribution of Thalassaemia Births in Pulau Pinang by Year

12.2.1 Age Groups

The youngest patient in Pulau Pinang is one year and 4 months old with β -thalassaemia intermedia and the eldest is 88 years old with Hb H disease. A total of 320 patients (61.19%) are below 20 years old.

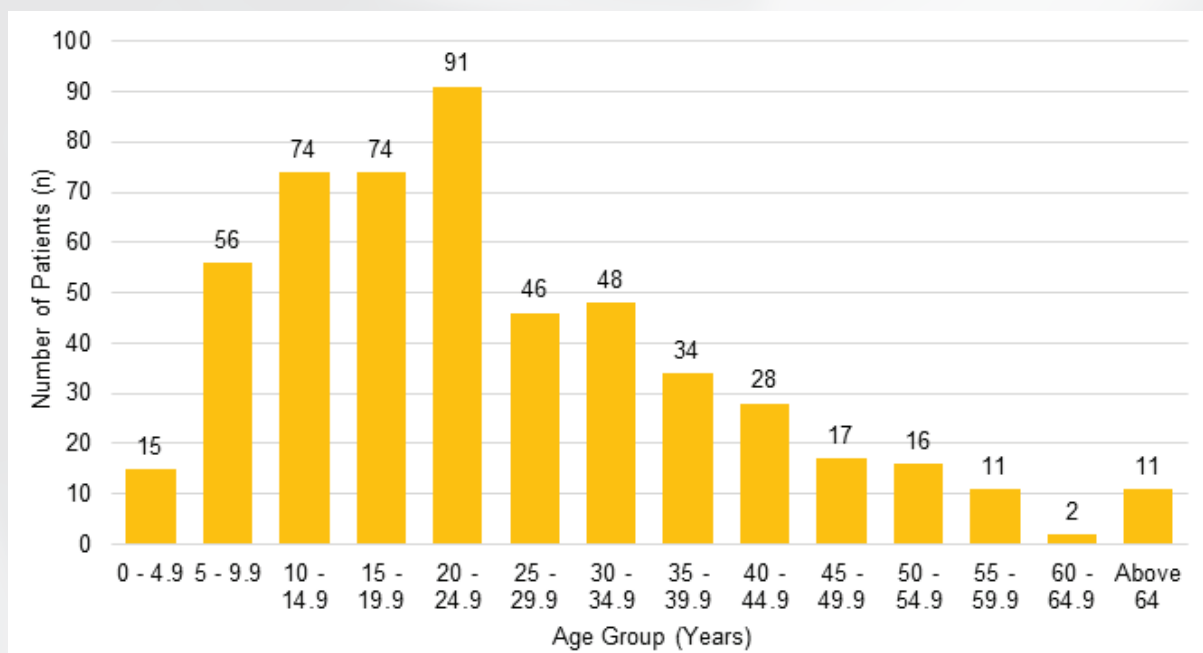


Figure 12.2: Distribution of Patients in Pulau Pinang by Age Group

Table 12.3: Distribution of Patients in Pulau Pinang According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0-14.9	145	β-Thalassaemia Major	29	20.00
		β-Thalassaemia Intermedia	15	10.34
		HbE/β-Thalassaemia	58	40.00
		Hb H Disease	35	24.14
		Others	8	5.52
15-29.9	211	β-Thalassaemia Major	54	25.59
		β-Thalassaemia Intermedia	12	5.69
		HbE/β-Thalassaemia	89	42.18
		Hb H Disease	53	25.12
		Others	3	1.42
30-44.9	110	β-Thalassaemia Major	23	20.91
		β-Thalassaemia Intermedia	12	10.91
		HbE/β-Thalassaemia	47	42.73
		Hb H Disease	22	20.00
		Others	6	5.45
45-59.9	44	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	5	11.36
		HbE/β-Thalassaemia	13	29.55
		Hb H Disease	24	54.55
		Others	2	4.55
60 and above	13	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	3	23.08
		Hb H Disease	10	76.92
		Others	0	0.00
Total			523	

12.2.2 Gender

There are 229 male patients (43.79%) and 294 female patients (56.21%) in Pulau Pinang.

Table 12.4: Distribution of Patients in Pulau Pinang According to Gender by Centre

Centre	Total Number of Patients	Male		Female	
		No.	%	No.	%
Hospital Pulau Pinang	282	120	22.94	162	30.98
Hospital Seberang Jaya	241	109	20.84	132	25.24
Total	523	229	43.79	294	56.21

12.2.3 Ethnic Group

The Malay form the largest group of patients in Pulau Pinang with 412 patients (78.78%), followed by Chinese with 95 patients (18.16%), Indian with four patients (0.76%), Kadazan Dusun with two patients (0.38%). The remaining 10 (1.19%) patients included Iban, foreigner and Thai descent.

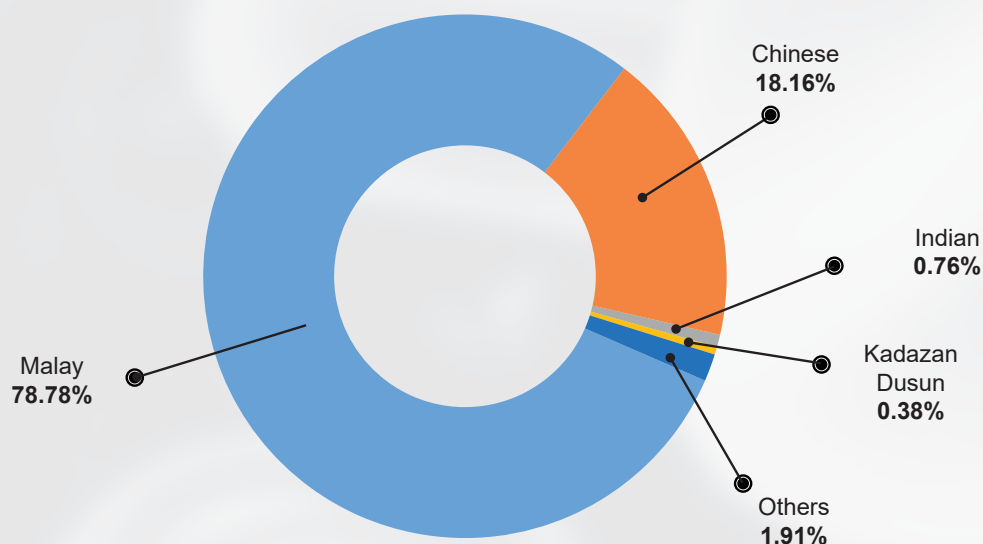


Figure 12.3: Distribution of Patients in Pulau Pinang by Ethnic Group

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

Centre	Total Number of Patients	Malay		Chinese		Indian		Kadazan Dusun		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Pulau Pinang	282	195	37.28	77	14.72	1	0.19	2	0.38	7	1.34
Hospital Seberang Jaya	241	217	41.49	18	3.44	3	0.57	0	0.00	3	0.57
Total	523	412	78.78	95	18.16	4	0.76	2	0.38	10	1.91

12.3 DIAGNOSIS

As shown in Figure 12.4, The most common diagnosis among patients in Pulau Pinang is HbE/ β -thalassaemia with 210 patients (40.15%), followed by Hb H disease with 144 patients (27.53%), β -thalassaemia major with 106 patients (20.27%), β -thalassaemia intermedia with 44 patient (8.41%). The remaining 19 patients (3.63%) consist of other diagnoses.

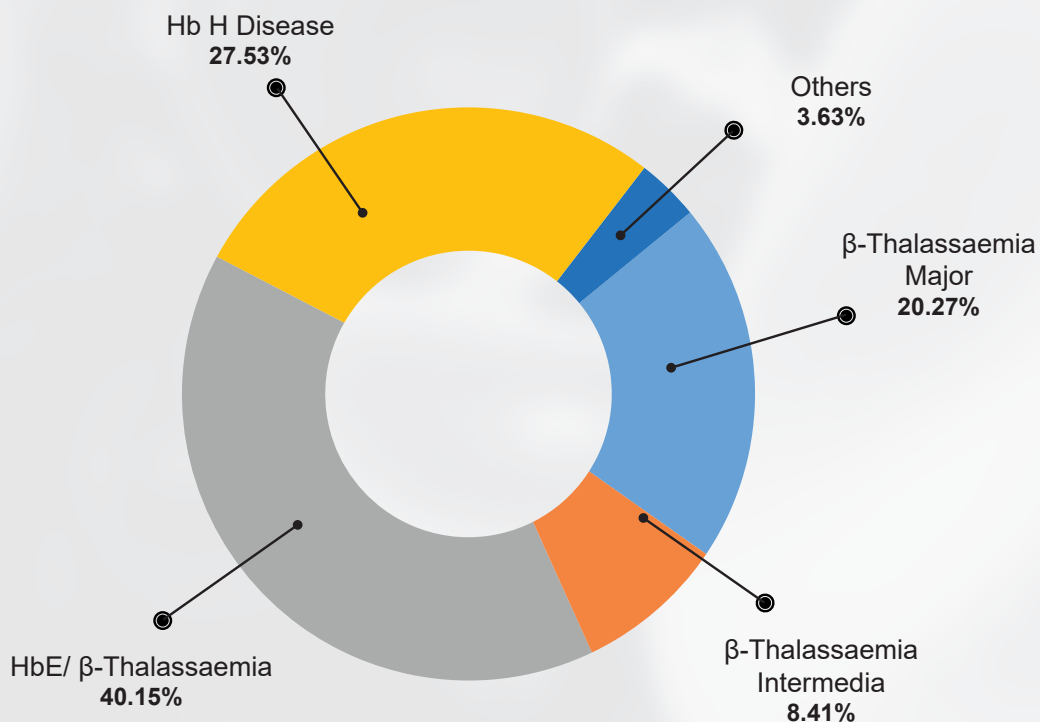


Figure 12.4: Distribution of Patients in Pulau Pinang by Diagnosis

Table 12.6: 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	106	Malay	73	13.96
		Chinese	30	5.74
		Indian	1	0.19
		Kadazan Dusun	1	0.19
		Others	1	0.19
β-Thalassaemia Intermedia	44	Malay	33	6.31
		Chinese	8	1.53
		Indian	1	0.19
		Kadazan Dusun	1	0.19
		Others	1	0.19
HbE/β-Thalassaemia	210	Malay	190	36.33
		Chinese	14	2.68
		Indian	2	0.38
		Kadazan Dusun	0	0.00
		Others	4	0.76
Hb H Disease	144	Malay	101	19.31
		Chinese	39	7.46
		Indian	0	0.00
		Kadazan Dusun	0	0.00
		Others	4	0.76
Others	19	Malay	15	2.87
		Chinese	3	0.57
		Indian	1	0.19
		Kadazan Dusun	0	0.00
		Others	0	0.00
Total			523	100.00

12.4 TREATMENT

12.4.1 Iron Chelation Therapy

A total of 326 (62.33%) out of 523 patients received iron chelation therapy. Patients who received monotherapy are 107 patients (32.82%) prescribed DFP, 71 patients (21.78%) prescribed DFX and 66 patients (20.25%) prescribed DFO. For a combination therapy, there are 62 patients (19.02%) prescribed DFO + DFP, 11 patients (3.37%) received DFO + DFX and nine patients (2.76%) prescribed DFP + DFX.

Table 12.7: Distribution of Patients in Pulau Pinang According by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	66	20.25
DFP only	107	32.82
DFX only	71	21.78
DFO + DFP	62	19.02
DFP + DFX	9	2.76
DFO + DFX	11	3.37
DFO + DFP + DFX	0	0.00
Total	326	100.00

Table 12.8: 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	155	DFO only	26	7.98
		DFP only	48	14.72
		DFX only	35	10.74
		DFO + DFP	37	11.35
		DFP + DFX	5	1.53
		DFO + DFX	4	1.23
		DFO + DFP + DFX	0	0.00
Hospital Seberang Jaya	177	DFO only	40	12.27
		DFP only	59	18.10
		DFX only	36	11.04
		DFO + DFP	25	7.67
		DFP + DFX	4	1.23
		DFO + DFX	7	2.15
		DFO + DFP + DFX	0	0.00
Total			326	100.00

Table 12.9: 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	83	DFO only	16	19.28
		DFP only	17	20.48
		DFX only	36	43.37
		DFO + DFP	10	12.05
		DFP + DFX	1	1.20
		DFO + DFX	3	3.61
		DFO + DFP + DFX	0	0.00
15-29.9	140	DFO only	23	16.43
		DFP only	39	27.86
		DFX only	30	21.43
		DFO + DFP	37	26.43
		DFP + DFX	6	4.29
		DFO + DFX	5	3.57
		DFO + DFP + DFX	0	0.00
30-44.9	74	DFO only	17	22.97
		DFP only	40	54.05
		DFX only	3	4.05
		DFO + DFP	11	14.86
		DFP + DFX	2	2.70
		DFO + DFX	1	1.35
		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			326	

12.4.2 Serum Ferritin Level

There are 148 regularly transfused patients in Pulau Pinang who had their serum ferritin levels measured in 2020. Approximately 43 patients (29.05%) had serum ferritin level below than 1,000 ng/mL, 45 patients (30.41%) have serum ferritin level between 1,000 and 2,500 ng/mL, and 60 patients (40.55%) have a serum ferritin level above 2,500 ng/mL.

Table 12.10: Distribution of TDT 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	126	38	25.85	39	25.85	32	21.77	14	9.52	3	2.04
Hospital Seberang Jaya	22	5	3.40	6	4.08	6	4.08	3	2.04	2	1.36
Total	148	43	29.05	45	30.41	38	25.68	17	11.49	5	3.38

12.5 COMPLICATIONS AND DEATHS

12.5.1 Complications

Hepatitis B, Hepatitis C and HIV are screened for all thalassaemia patients. There are 13 patients who are reported with anti-HCV positive. However, all these patients are HCV RNA negative. This is determined as past exposure infection with no recent active infection and seven patients were successfully treated for Hepatitis C infection previously.

Table 12.11: Distribution of Patients in Pulau Pinang According to Tranfusion Transmissible Infections

Complications	2007-2019	2020
Hepatitis B	0	0
Hepatitis C: Anti HCV	13	0
HCV RNA	0	0
HIV	1	0
Total	13	0

Based on Table 12.12, there are five patients with delayed puberty, five patients with short stature, four patients with hypothyroid and three patients are on treatment for diabetes mellitus.

Table 12.12: Distribution of Patients in Pulau Pinang According to Endocrine Complications

Complications	Number of Patients (n)
Delayed Puberty	5
Short Stature	5
Hypothyroid	4
Diabetes Mellitus	3
Total	17

12.5.2 Iron Deposition in Heart and Liver

The liver and cardiac MRI T2* scan will be done yearly. The severity of cardiac and liver iron loading of patients were recorded as shown in Tables 12.13 and 12.14.

There are 68 patients from Hospital Pulau Pinang who had their latest liver and cardiac MRI T2* scan done. Sixty patients (88.24%) have no cardiac iron deposition. Fifteen patients (22.06%) had normal liver iron deposition. Patients with severe iron loading are treated with intensive chelation.

Table 12.13: Distribution of Patients in Pulau Pinang According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Pulau Pinang	68	60	88.24	2	2.94	0	0.00	6	8.82
Total	68	60	88.24	2	2.94	0	0.00	6	8.82

Table 12.14: Distribution of Patients in Pulau Pinang According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Pulau Pinang	68	15	22.06	22	32.35	11	16.18	20	29.41
Total	68	15	22.06	22	32.35	11	16.18	20	29.41

12.5.3 Deaths

Based on Table 12.15, there were 18 cumulative deaths for thalassaemia patients reported. In 2020, two patients died. A patient with β -thalassaemia major passed away due to cardiac while the other patient with HbE/ β -thalassaemia died due to infection.

Table 12.15: Cumulative Known Causes of Death in Pulau Pinang

Causes of Death	Number of Patients (n)
Infections	8
Cardiac	4
Renal Complications	2
Central Nervous System Event	2
Malignancy	1
Motor Vehicle Accident (MVA)	1
Total	18

12.6 CONCLUSION

The most prevalent type of thalassemia in Pulau Pinang is HbE/ β -thalassaemia. The median patient age group is 21-25 years old. In Pulau Pinang, there are 13 patients with positive anti HCV. However, all of them are HCV RNA negative. This indicates either past exposure to Hepatitis C or successful treatment.

About 88 TDT patients (59.46%) achieved serum ferritin level below 2,499 ng/mL. 60 patients (40.54%) were still inadequately chelated with serum ferritin level above 2,500 ng/mL.

The most common endocrine complication among the patients are delayed puberty and short stature. Out of the 68 patients who had liver and cardiac MRI T2* scan done, 8.82% patients have severe cardiac iron overload and 20 patients (29.41%) has severe liver iron overload.



13.0
SABAH

13.1 INTRODUCTION

Sabah is the second largest state in Malaysia after Sarawak. The population of Sabah in 2020 was reported at 3,908,500 (Department of Statistics Malaysia).

The population in 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.

13.2 PATIENT DEMOGRAPHICS

There are 1,907 thalassaemia patients in Sabah registered in the MTR until December 2020.

Table 13.1: Distribution of Patients in Sabah by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Queen Elizabeth	372	19.51
Hospital Wanita & Kanak-Kanak Sabah	320	16.78
Hospital Duchess of Kent	201	10.54
Hospital Keningau	191	10.02
Hospital Kota Marudu	146	7.66
Hospital Ranau	97	5.09
Hospital Pitas	93	4.88
Hospital Kudat	79	4.14
Hospital Kota Belud	59	3.09
Hospital Lahad Datu	50	2.62
Hospital Tawau	46	2.41
Hospital Papar	45	2.36
Hospital Tambunan	37	1.94
Hospital Kinabatangan	29	1.52
Hospital Beaufort	28	1.47
Hospital Tenom	24	1.26
Hospital Tuaran	22	1.15
Hospital Sipitang	20	1.05
Hospital Beluran	18	0.94
Hospital Kuala Penyu	17	0.89
Hospital Kunak	11	0.58
Hospital Semporna	2	0.10
Total	1907	100.00

There are three hospitals with the highest number of patients in Sabah namely Hospital Queen Elizabeth with 372 patients (19.51%), Hospital Wanita & Kanak-Kanak Sabah, Kota Kinabalu with 320 patients (16.78%) and Hospital Duchess of Kent with 201 patients (10.54%).

The ethnic groups have a significant impact on thalassaemia carrier rate in Sabah. β -thalassaemia are more common in Kadazan Dusun, Rungus, Bajau and Murut. Hence, this explains why small districts, such as Keningau, Kota Marudu, Pitas and Ranau have a large number of thalassaemia patients. The hospitals in the Southeast of Sabah, such as Tawau and Lahad Datu Hospitals have relatively fewer patients as there is relatively low density of Kadazan Dusun, Rungus, Bajau and Murut populations in these towns.

Table 13.2: Distribution of Patients in Sabah by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	1747
Cured by Stem Cell Therapy	39
Total	1786
Lost to Follow-up	121
Total	1907
Deaths in 2020	24
Cumulative Reported Deaths	349

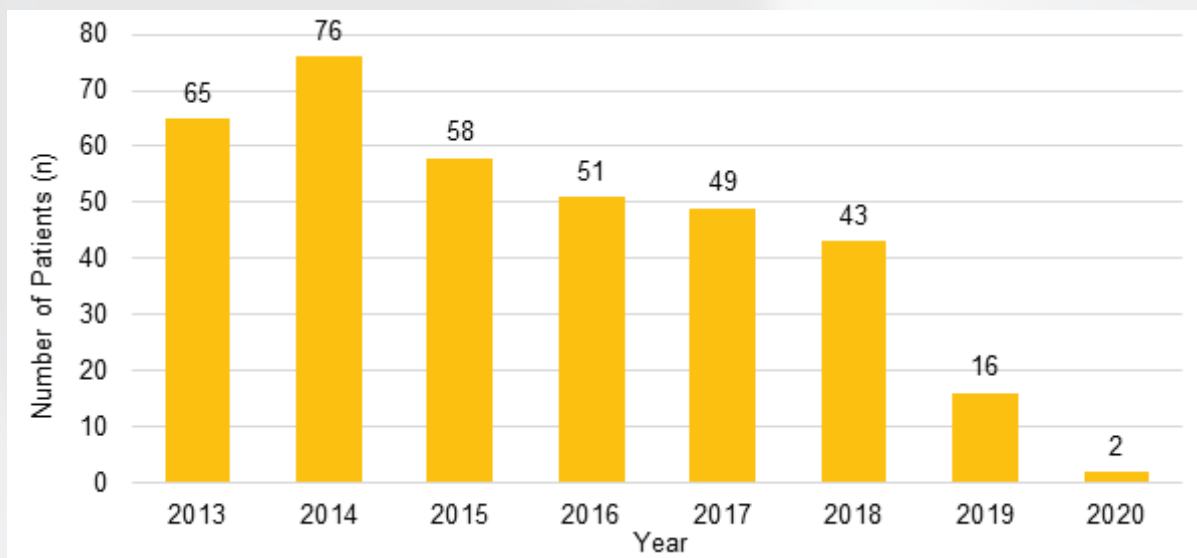


Figure 13.1: Distribution of Thalassaemia Births in Sabah by Year

13.2.1 Age Groups

The age group with the highest number of thalassaemia patients in Sabah is 10-14.9 years. The oldest patient surviving with β -Thalassaemia Major in Sabah is currently 50 years old. In 2020, the number of thalassaemia patients younger than 15 years old continues to increase. However, the overall percentage of patients younger than 15 years old in 2020 has reduced in view of increasing survival rates of whole cohorts of thalassaemia patients.

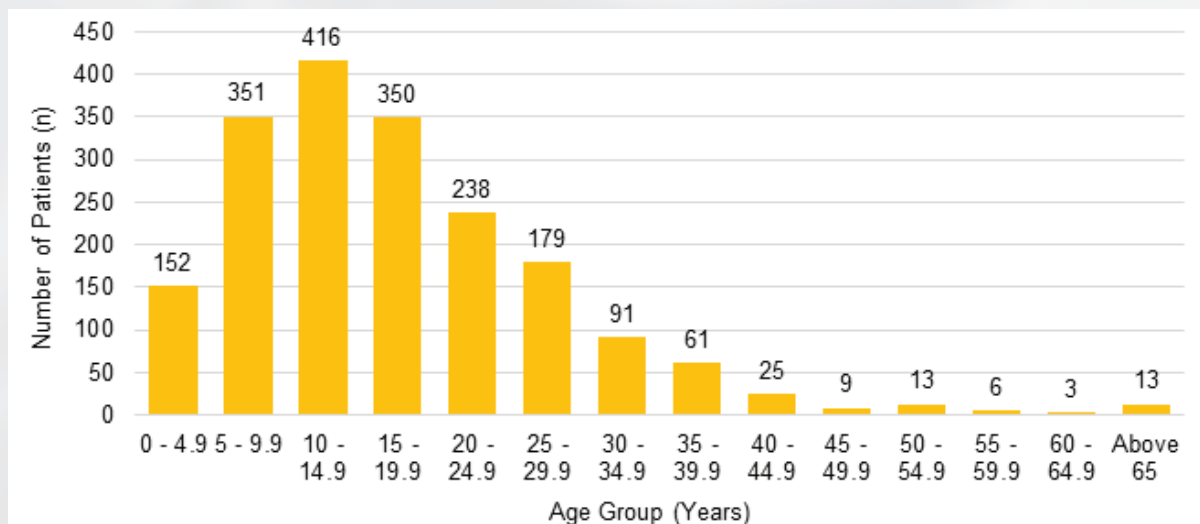


Figure 13.2: Distribution of Patients in Sabah by Age Group

Table 13.3: Distribution of Patients in Sabah According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0-14.9	919	β -Thalassaemia Major	677	73.67
		β -Thalassaemia Intermedia	104	11.32
		HbE/ β -Thalassaemia	76	8.27
		Hb H Disease	62	6.75
		Others	0	0.00
15-29.9	767	β -Thalassaemia Major	590	76.92
		β -Thalassaemia Intermedia	119	15.51
		HbE/ β -Thalassaemia	39	5.08
		Hb H Disease	19	2.48
		Others	0	0.00
30-44.9	177	β -Thalassaemia Major	76	42.94
		β -Thalassaemia Intermedia	48	27.12
		HbE/ β -Thalassaemia	25	14.12
		Hb H Disease	28	15.82
		Others	0	0.00
45-59.9	28	β -Thalassaemia Major	2	7.14
		β -Thalassaemia Intermedia	6	21.43
		HbE/ β -Thalassaemia	11	39.29
		Hb H Disease	9	32.14
		Others	0	0.00
60 and above	16	β -Thalassaemia Major	0	0.00
		β -Thalassaemia Intermedia	2	12.50
		HbE/ β -Thalassaemia	2	12.50
		Hb H Disease	12	75.00
		Others	0	0.00
Total			1907	

β -thalassaemia major constitutes the majority diagnosis in Sabah. Until December 2020, 70.52% of total number of patients were diagnosed with β -thalassaemia major, and 14.63% 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 finding 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.

13.2.2 Gender

The gender distribution of thalassaemia patients in Sabah is consistent with thalassaemia syndrome as an autosomal recessive disease.

Table 13.4: Distribution of Patients in Sabah According to Gender by Centre

Centre	Total Number of Patients	Male		Female	
		No.	%	No.	%
Hospital Queen Elizabeth	372	172	9.02	200	10.49
Hospital Wanita & Kanak-Kanak Sabah	320	166	8.70	154	8.08
Hospital Dutchess of Kent	201	101	5.30	100	5.24
Hospital Keningau	191	101	5.30	90	4.72
Hospital Kota Marudu	146	75	3.93	71	3.72
Hospital Ranau	97	57	2.99	40	2.10
Hospital Pitas	93	49	2.57	44	2.31
Hospital Kudat	79	40	2.10	39	2.05
Hospital Kota Belud	59	32	1.68	27	1.42
Hospital Lahad Datu	50	21	1.10	29	1.52
Hospital Tawau	46	22	1.15	24	1.26
Hospital Papar	45	34	1.78	11	0.58
Hospital Tambunan	37	22	1.15	15	0.79
Hospital Kinabatangan	29	14	0.73	15	0.79
Hospital Beaufort	28	18	0.94	10	0.52
Hospital Tenom	24	11	0.58	13	0.68
Hospital Tuaran	22	15	0.79	7	0.37
Hospital Sipitang	20	14	0.73	6	0.31
Hospital Beluran	18	11	0.58	7	0.37
Hospital Kuala Penyu	17	12	0.63	5	0.26
Hospital Kunak	11	6	0.31	5	0.26
Hospital Semporna	2	0	0.00	2	0.10
Total	1907	993	52.07	914	47.93

13.2.3 Ethnic Group

The Kadazan Dusun form the largest group of patients in Sabah with 897 patients (47.04%), followed by Pribumi Sabah with 244 patients (12.79%).

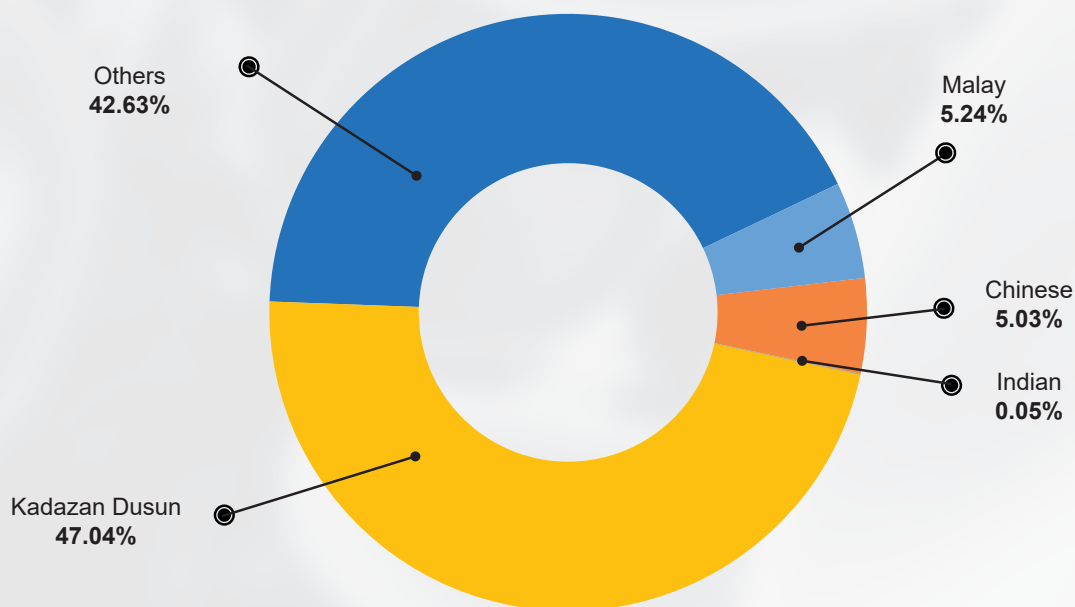


Figure 13.3: Distribution of Patients in Sabah by Ethnic Group

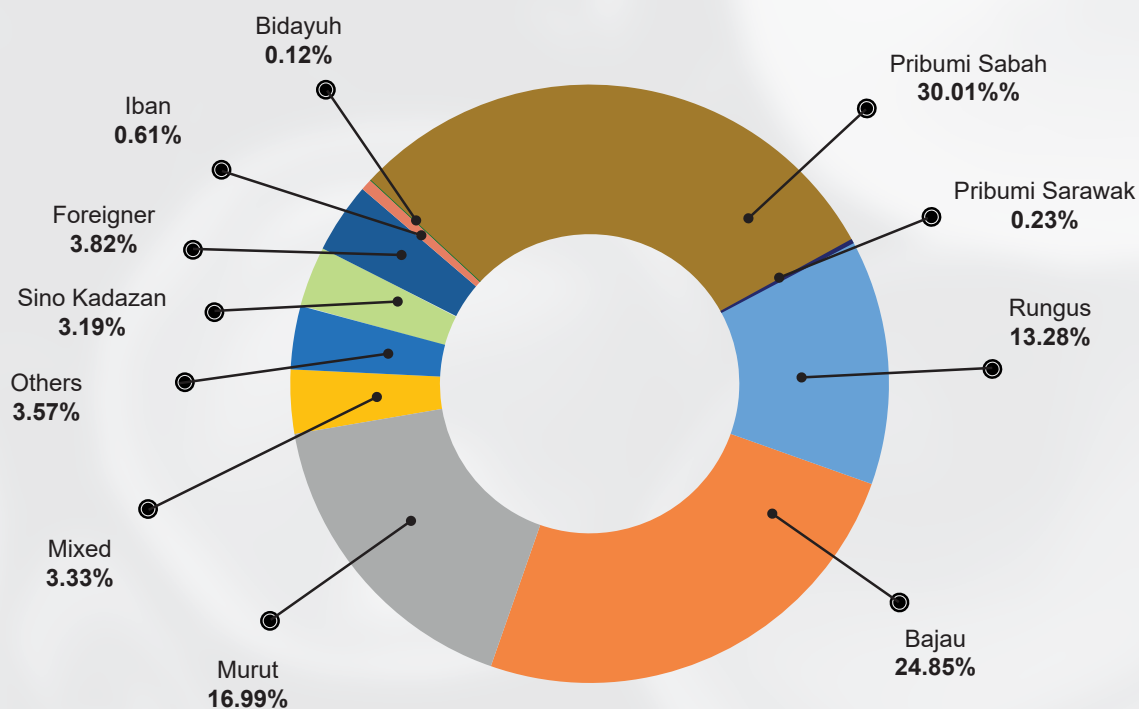


Figure 13.4: Distribution of Patients in Sabah by Ethnic Sub-Group of Others

Table 13.5: Distribution of Patients in Sabah According to Ethnic Group by Centre

Centre	Total Number of Patients	Kadazan Dusun		Rungus		Malay		Bajau		Murut		Chinese		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Queen Elizabeth	372	167	8.76	25	1.31	26	1.36	56	2.94	9	0.47	47	2.46	42	2.20
Hospital Wanita & Kanak-kanak Sabah	320	158	8.29	12	0.63	20	1.05	39	2.05	10	0.52	19	1.00	62	3.25
Hospital Dutchess of Kent	201	57	2.99	0	0.00	16	0.84	25	1.31	0	0.00	7	0.37	96	5.03
Hospital Keningau	191	94	4.93	0	0.00	1	0.05	1	0.05	83	4.35	6	0.31	6	0.31
Hospital Kota Marudu	146	108	5.66	10	0.52	2	0.10	6	0.31	0	0.00	1	0.05	19	1.00
Hospital Ranau	97	97	5.09	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Pitas	93	46	2.41	13	0.68	1	0.05	1	0.05	0	0.00	1	0.05	31	1.63
Hospital Kudat	79	10	0.52	45	2.36	1	0.05	8	0.42	0	0.00	4	0.21	11	0.58
Hospital Kota Belud	59	39	2.05	1	0.05	3	0.16	11	0.58	1	0.05	0	0.00	4	0.21
Hospital Lahad Datu	50	6	0.31	0	0.00	4	0.21	20	1.05	0	0.00	2	0.10	18	0.94
Hospital Tawau	46	4	0.21	0	0.00	6	0.31	12	0.63	0	0.00	6	0.31	18	0.94
Hospital Papar	45	23	1.21	2	0.10	7	0.37	5	0.26	3	0.16	0	0.00	5	0.26
Hospital Tambunan	37	33	1.73	0	0.00	0	0.00	0	0.00	2	0.10	0	0.00	2	0.10
Hospital Kinabatangan	29	0	0.00	0	0.00	3	0.16	2	0.10	0	0.00	0	0.00	24	1.26

Hospital Beaufort	28	5	0.26	0	0.00	4	0.21	2	0.10	2	0.10	0	0.00	15	0.79
Hospital Tenom	24	2	0.10	0	0.00	2	0.10	1	0.05	16	0.84	2	0.10	1	0.05
Hospital Tuaran	22	19	1.00	0	0.00	0	0.00	1	0.05	0	0.00	1	0.05	1	0.05
Hospital Sipitang	20	2	0.10	0	0.00	3	0.16	1	0.05	12	0.63	0	0.00	2	0.10
Hospital Beluran	18	14	0.73	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	4	0.21
Hospital Kuala Penyu	17	10	0.52	0	0.00	1	0.05	3	0.16	0	0.00	0	0.00	3	0.16
Hospital Kunak	11	3	0.16	0	0.00	0	0.00	6	0.31	0	0.00	0	0.00	2	0.10
Hospital Semporna	2	0	0.00	0	0.00	0	0.00	2	0.10	0	0.00	0	0.00	0	0.00
Total	1907	897	47.04	108	5.66	100	5.24	202	10.59	138	7.24	96	5.03	366	19.19

A significant percentage of thalassaemia patients in Sabah are of Kadazan Dusun descent (47.04%) 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 case is also disproportionately high among the Bajau (10.59%) and Murut (7.49%).

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. There are 70.53% of thalassaemia patients in Sabah were diagnosed with β -thalassaemia major and 14.63% patients were diagnosed with β -thalassaemia intermedia. HbE/ β -thalassaemia constitutes much fewer cases in Sabah as compared to West Malaysia. This observation is consistent with the discovery of the severe mutation such as 45-kb β -globin gene deletion (Filipino deletion), occurring in homozygous form in more than 90% of Sabahan Kadazan Dusun with β -thalassaemia syndrome.

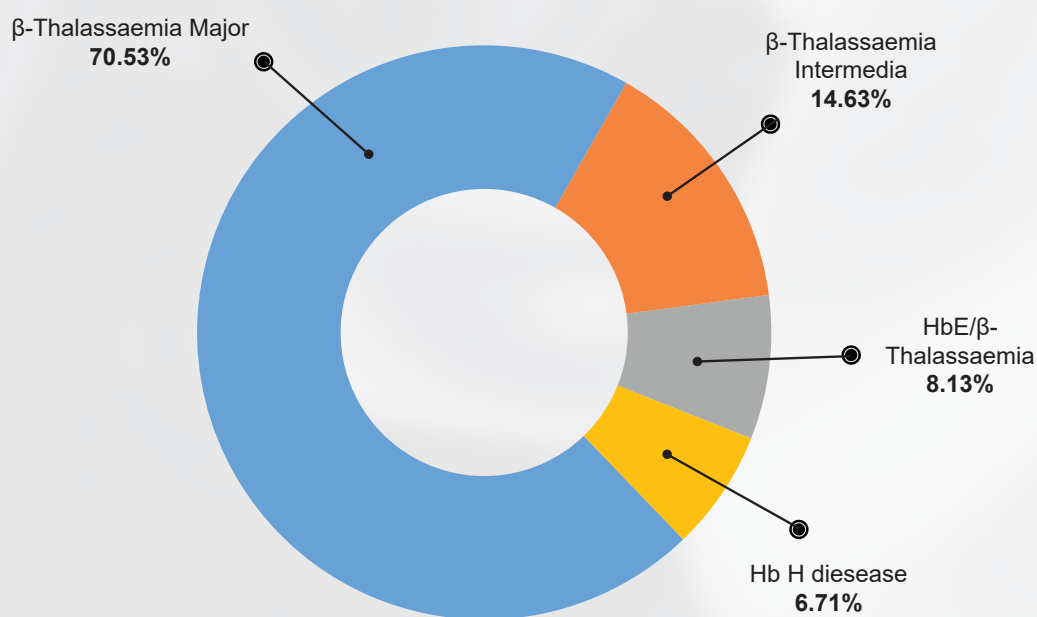


Figure 13.5: Distribution of Patients in Sabah by Diagnosis

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

Centre	Total Number of Patients	β-Thalassaemia major		β-Thalassaemia intermedia		HbE/β-Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Queen Elizabeth	372	219	11.48	60	3.15	42	2.20	51	2.67	0	0.00
Hospital Wanita & Kanak-Kanak Sabah	320	210	11.01	58	3.04	27	1.42	25	1.31	0	0.00
Hospital Dutchess of Ken	201	113	5.93	46	2.41	33	1.73	9	0.47	0	0.00
Hospital Keningau	191	172	9.02	13	0.68	4	0.21	2	0.10	0	0.00
Hospital Kota Marudu	146	120	6.29	18	0.94	6	0.31	2	0.10	0	0.00
Hospital Ranau	97	88	4.61	7	0.37	2	0.10	0	0.00	0	0.00
Hospital Pitas	93	76	3.99	7	0.37	7	0.37	3	0.16	0	0.00
Hospital Kudat	79	52	2.73	4	0.21	8	0.42	15	0.79	0	0.00
Hospital Kota Belud	59	40	2.10	8	0.42	9	0.47	2	0.10	0	0.00
Hospital Lahad Datu	50	25	1.31	9	0.47	6	0.31	10	0.52	0	0.00
Hospital Tawau	46	22	1.15	12	0.63	6	0.31	6	0.31	0	0.00
Hospital Papar	45	37	1.94	6	0.31	2	0.10	0	0.00	0	0.00
Hospital Tambunan	37	35	1.84	2	0.10	0	0.00	0	0.00	0	0.00
Hospital Kinabatangan	29	24	1.26	3	0.16	2	0.10	0	0.00	0	0.00

Hospital Beaufort	28	21	1.10	7	0.37	0	0.00	0	0.00	0	0.00
Hospital Tenom	24	21	1.10	1	0.05	1	0.05	1	0.05	0	0.00
Hospital Tuaran	22	20	1.05	2	0.10	0	0.00	0	0.00	0	0.00
Hospital Sipitang	20	16	0.84	3	0.16	0	0.00	1	0.05	0	0.00
Hospital Beluran	18	18	0.94	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kuala Penyu	17	10	0.52	6	0.31	0	0.00	1	0.05	0	0.00
Hospital Kunak	11	4	0.21	7	0.37	0	0.00	0	0.00	0	0.00
Hospital Semporna	2	2	0.10	0	0.00	0	0.00	0	0.00	0	0.00
Total	1907	1345	70.53	279	14.63	155	8.13	128	6.71	0	0.00

Table 13.7: 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	1345	Malay	51	2.67
		Chinese	46	2.41
		Indian	1	0.05
		Kadazan Dusun	770	40.38
		Others	477	25.01
β-Thalassaemia Intermedia	279	Malay	22	1.15
		Chinese	8	0.42
		Indian	0	0.00
		Kadazan Dusun	88	4.61
		Others	161	8.44
HbE/β-Thalassaemia	155	Malay	18	0.94
		Chinese	6	0.31
		Indian	0	0.00
		Kadazan Dusun	24	1.26
		Others	107	5.61
Hb H Disease	128	Malay	9	0.47
		Chinese	36	1.89
		Indian	0	0.00
		Kadazan Dusun	15	0.79
		Others	68	3.57
Others	0	Malay	0	0.00
		Chinese	0	0.00
		Indian	0	0.00
		Kadazan Dusun	0	0.00
		Others	0	0.00
Total			1907	100.00

13.4 TREATMENT

13.4.1 Iron Chelation Therapy

Table 13.8 shows that the combination of DFO and DFP is the most common type of iron chelator prescribed (25.63%). Meanwhile, DFX is mainly prescribed to paediatric patients. Most often older thalassaemia major patients in Sabah started chelation therapy later in life, such as when the government provided free chelation treatment beginning in 2006. The majority of these patients already had severe iron loading, hence these patients are treated with a combination of DFO and DFP. A total of 553 patients in Sabah were not prescribed with iron chelation therapy.

Table 13.8: Distribution of Patients in Sabah by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	274	20.24
DFP only	271	20.01
DFX only	311	22.97
DFO + DFP	347	25.63
DFP + DFX	76	5.61
DFO + DFX	49	3.62
DFO + DFP + DFX	26	1.92
Total	1354	100.00

Table 13.9: 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	246	DFO only	111	8.20
		DFP only	33	2.44
		DFX only	3	0.22
		DFO + DFP	75	5.54
		DFP + DFX	1	0.07
		DFO + DFX	6	0.44
		DFO + DFP + DFX	17	1.26
Hospital Wanita & Kanak-kanak Sabah	192	DFO only	16	1.18
		DFP only	45	3.32
		DFX only	76	5.61
		DFO + DFP	35	2.58
		DFP + DFX	10	0.74
		DFO + DFX	10	0.74
		DFO + DFP + DFX	0	0.00
Hospital Dutchess of Kent	153	DFO only	33	2.44
		DFP only	53	3.91
		DFX only	27	1.99
		DFO + DFP	32	2.36
		DFP + DFX	3	0.22
		DFO + DFX	4	0.30
		DFO + DFP + DFX	1	0.07
Hospital Keningau	151	DFO only	14	1.03
		DFP only	29	2.14
		DFX only	32	2.36
		DFO + DFP	45	3.32
		DFP + DFX	21	1.55
		DFO + DFX	7	0.52
		DFO + DFP + DFX	3	0.22
Hospital Kota Marudu	112	DFO only	5	0.37
		DFP only	38	2.81
		DFX only	15	1.11
		DFO + DFP	43	3.18
		DFP + DFX	10	0.74
		DFO + DFX	1	0.07
		DFO + DFP + DFX	0	0.00
Hospital Pitas	68	DFO only	11	0.81
		DFP only	14	1.03
		DFX only	23	1.70
		DFO + DFP	16	1.18
		DFP + DFX	4	0.30
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Ranau	61	DFO only	31	2.29
		DFP only	2	0.15
		DFX only	20	1.48
		DFO + DFP	6	0.44
		DFP + DFX	0	0.00
		DFO + DFX	1	0.07
		DFO + DFP + DFX	1	0.07
Hospital Kudat	59	DFO only	11	0.81
		DFP only	6	0.44
		DFX only	20	1.48
		DFO + DFP	19	1.40
		DFP + DFX	1	0.07
		DFO + DFX	1	0.07
		DFO + DFP + DFX	1	0.07
Hospital Kota Belud	42	DFO only	6	0.44
		DFP only	6	0.44
		DFX only	13	0.96
		DFO + DFP	15	1.11
		DFP + DFX	1	0.07
		DFO + DFX	1	0.07
		DFO + DFP + DFX	0	0.00
Hospital Lahad Datu	34	DFO only	4	0.30
		DFP only	4	0.30
		DFX only	12	0.89
		DFO + DFP	12	0.89
		DFP + DFX	1	0.07
		DFO + DFX	1	0.07
		DFO + DFP + DFX	0	0.00
Hospital Tawau	33	DFO only	1	0.07
		DFP only	4	0.30
		DFX only	10	0.74
		DFO + DFP	7	0.52
		DFP + DFX	4	0.30
		DFO + DFX	6	0.44
		DFO + DFP + DFX	1	0.07
Hospital Tambunan	33	DFO only	4	0.30
		DFP only	1	0.07
		DFX only	9	0.66
		DFO + DFP	7	0.52
		DFP + DFX	9	0.66
		DFO + DFX	3	0.22
		DFO + DFP + DFX	0	0.00
Hospital Papar	30	DFO only	4	0.30
		DFP only	7	0.52
		DFX only	12	0.89

		DFO + DFP	2	0.15
		DFP + DFX	2	0.15
		DFO + DFX	3	0.22
		DFO + DFP + DFX	0	0.00
Hospital Kinabatangan	24	DFO only	7	0.52
		DFP only	7	0.52
		DFX only	4	0.30
		DFO + DFP	5	0.37
		DFP + DFX	1	0.07
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Beaufort	23	DFO only	4	0.30
		DFP only	3	0.22
		DFX only	10	0.74
		DFO + DFP	5	0.37
		DFP + DFX	0	0.00
		DFO + DFX	1	0.07
		DFO + DFP + DFX	0	0.00
Hospital Tenom	21	DFO only	5	0.37
		DFP only	7	0.52
		DFX only	1	0.07
		DFO + DFP	8	0.59
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tuaran	20	DFO only	0	0.00
		DFP only	3	0.22
		DFX only	8	0.59
		DFO + DFP	1	0.07
		DFP + DFX	8	0.59
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Sipitang	17	DFO only	1	0.07
		DFP only	2	0.15
		DFX only	9	0.66
		DFO + DFP	3	0.22
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	2	0.15
Hospital Beluran	16	DFO only	4	0.30
		DFP only	3	0.22
		DFX only	4	0.30
		DFO + DFP	5	0.37
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Kuala Penyu	10	DFO only	2	0.15
		DFP only	1	0.07
		DFX only	1	0.07
		DFO + DFP	2	0.15
		DFP + DFX	0	0.00
		DFO + DFX	4	0.30
		DFO + DFP + DFX	0	0.00
Hospital Kunak	7	DFO only	0	0.00
		DFP only	2	0.15
		DFX only	1	0.07
		DFO + DFP	4	0.30
		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	0.07
		DFX only	1	0.07
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			1354	100.00

Table 13.10: 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	621	DFO only	53	8.53
		DFP only	117	18.84
		DFX only	295	47.50
		DFO + DFP	60	9.66
		DFP + DFX	63	10.14
		DFO + DFX	29	4.67
		DFO + DFP + DFX	4	0.64
15-29.9	623	DFO only	195	31.30
		DFP only	116	18.62
		DFX only	16	2.57
		DFO + DFP	250	40.13
		DFP + DFX	12	1.93
		DFO + DFX	18	2.89
		DFO + DFP + DFX	16	2.57
30-44.9	100	DFO only	23	23.00
		DFP only	31	31.00
		DFX only	0	0.00
		DFO + DFP	37	37.00
		DFP + DFX	1	1.00
		DFO + DFX	2	2.00
		DFO + DFP + DFX	6	6.00
45-59.9	10	DFO only	3	30.00
		DFP only	7	70.00
		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			1354	

13.4.2 Serum Ferritin Level

There are 1,123 patients in Sabah who had their serum ferritin level measured in 2020. A total of 299 patients (26.63%) has a serum ferritin level below 1,000 ng/mL, 294 patients (26.18%) have a serum ferritin level between 1,000-2,499 ng/mL, and 259 patients (23.06%) with a serum ferritin level between 2,500-4,999 ng/mL. Patients with a serum ferritin level between 5,000-9,999 ng/mL are 206 patients (18.34%), whereas number of patients with a serum ferritin level above 10,000 ng/mL are 65 patients (5.79%).

Table 13.11: Distribution of TDT 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.	%
Hospital Wanita & Kanak-kanak Sabah	213	88	7.84	69	6.14	41	3.65	14	1.25	1	0.09
Hospital Queen Elizabeth	204	53	4.72	50	4.45	40	3.56	38	3.38	23	2.05
Hospital Dutchess of Kent	150	29	2.58	42	3.74	40	3.56	29	2.58	10	0.89
Hospital Kota Marudu	108	36	3.21	27	2.40	23	2.05	21	1.87	1	0.09
Hospital Keningau	64	16	1.42	17	1.51	9	0.80	15	1.34	7	0.62
Hospital Kudat	62	18	1.60	16	1.42	15	1.34	11	0.98	2	0.18
Hospital Kota Belud	53	20	1.78	12	1.07	13	1.16	8	0.71	0	0.00
Hospital Ranau	49	5	0.45	11	0.98	16	1.42	13	1.16	4	0.36
Hospital Pitas	42	4	0.36	13	1.16	13	1.16	11	0.98	1	0.09

Hospital Lahad Datu	36	9	0.80	4	0.36	7	0.62	10	0.89	6	0.53
Hospital Tambunan	32	5	0.45	8	0.71	9	0.80	8	0.71	2	0.18
Hospital Kinabatangan	27	1	0.09	4	0.36	9	0.80	10	0.89	3	0.27
Hospital Beaufort	18	5	0.45	4	0.36	4	0.36	4	0.36	1	0.09
Hospital Tuaran	18	3	0.27	8	0.71	4	0.36	2	0.18	1	0.09
Hospital Sipitang	15	1	0.09	3	0.27	5	0.45	4	0.36	2	0.18
Hospital Papar	13	0	0.00	2	0.18	6	0.53	5	0.45	0	0.00
Hospital Kuala Penyu	12	5	0.45	2	0.18	2	0.18	2	0.18	1	0.09
Hospital Tenom	6	0	0.00	2	0.18	3	0.27	1	0.09	0	0.00
Hospital Beluran	1	1	0.09	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kunak	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Semporna	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Tawau	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	1123	299	26.63	294	26.18	259	23.06	206	18.34	65	5.79

13.5 COMPLICATIONS AND DEATHS

13.5.1 Complications

Based on Table 13.12, there are eight patients cumulatively with transfusion transmissible infections, one patient had Hepatitis B, 5 patients had Hepatitis C, and 2 patients had HIV. Due to incomplete and difficulty of data retrievals, there are no data recorded for other complications.

Table 13.12: Distribution of Patients in Sabah According to Transfusion Transmissible Infections

Complications	2007-2019	2020
Hepatitis C: Anti HCV	5	0
HCV RNA	0	0
Hepatitis B	1	0
HIV	2	0
Total	8	0

13.5.2 Iron Deposition in Heart and Liver Complications

The following Tables 13.13 and 13.14 show a total of patients who underwent for liver and cardiac MRI T2* scan. Some patients have not done the liver MRI scan in Sabah. The data in the section could not be interpreted by centre due to technical issue.

Table 13.13 shows that there are 458 patients had normal iron loading and 122 patients had severe iron loading in cardiac. Based on Table 13.14, 85 patients had normal iron loading and 353 patients had severe iron loading in their liver. Most of the patients who had abnormal result both for cardiac and liver are mainly due to non-compliance with prescribed iron chelator.

Table 13.13: Distribution of Patients in Sabah According to Cardiac MRI T2*

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Sabah	687	458	66.67	46	6.70	61	8.88	122	17.76
Total	687	458	66.67	46	6.70	61	8.88	122	17.76

Table 13.14: Distribution of Patients in Sabah According to Liver MRI

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Sabah	647	85	13.14	98	15.15	111	17.16	353	54.56
Total	647	85	13.14	98	15.15	111	17.16	353	54.56

13.5.3 Deaths

Table 13.15 presents a cumulative total of 304 known deaths reported in Sabah. The two highest causes of death among thalassaemia patients in Sabah are cardiac with 133 patients and infections with 112 patients. There are also eight deaths with endocrine complication and eight died at home reported. Additionally, there are 45 cases with unknown cause of death in Sabah due to unavailable data and these patients were not included in this table.

Table 13.15: Cumulative Known Causes of Death in Sabah

Causes of Death	Number of Patients (n)
Cardiac	133
Infections	112
Endocrine Complications	8
Died at Home/Brought in Dead to Hospital	8
Others	7
Liver Disease	7
Central Nervous System Event	7
Motor Vehicle Accident (MVA)	5
Thalasaemia	4
Malignancy	4
Surgical Complications	3
Renal Complications	2
Thrombosis	2
Bone Marrow Transplant Complications	1
Acute Haemolysis	1
Total	304

13.6 CONCLUSION

There are 93 new thalassaemia patients in Sabah for 2020. The highest number of thalassaemia patients are from Hospital Queen Elizabeth with 372 patients, followed by Hospital Wanita & Kanak-Kanak Sabah with 320 patients, Hospital Duchess of Kent with 210 patients and Hospital Keningau with 191 patients. In addition, 213 patients in Hospital Kota Marudu and 242 patients in Hospital Pitas were reported for highest notification rate of 100,000 population in 2020. It indicates that the density of thalassaemia patients over populations in the west Sabah is abundant than the other areas in Sabah.

Currently in Sabah, there are 993 male patients (52.07%) and 914 female patients (47.93%). Kadazan Dusun forms the largest ethnic group with 897 patients (47.04%) followed by Pribumi Sabah with 244 patients (12.79%) and Bajau with 202 patients (10.59%).

The highest total number of patients based on age group are 0-14.9 years old by 919 patients, followed by 15-29.9 years old by 767 patients and 30–44.9 years old by 177 patients. There are 70.53% of Sabah thalassaemia patients are diagnosed with β -thalassaemia major, followed by β -Thalassaemia intermedia with 14.63% patients, HbE/ β -thalassaemia with 8.13% patients and Hb H disease with 6.71% patients.

The total number of 1,354 patients are prescribed with iron chelation therapy. There are 856 patients received monotherapy, 472 patients are prescribed to a combination therapy and 26 patients are prescribed to all three chelators. The most common chelators among Sabah's Thalassaemia patients are combination of DFO+DFP and age group that are most prescribed with Iron chelators is age between 15-29.9 by 623 patients.

The number of patients who had their serum ferritin level measured in 2020 were only 1,123 patients. 299 patients (26.63%) have a serum ferritin level below 1,000 ng/mL whereas number of patients with serum ferritin level more than 10,000 ng/ml are 65 patients (5.40%).

There were 8 patients have been confirmed with transfusion transmissible infections. 5 patients had Hepatitis C, two patients had HIV and one patient had Hepatitis B. There are no records for other complications among Sabah patients. Until 2020, there are 349 cumulative deaths included 45 cases with unknown cause of death were reported in Sabah. The highest death with 133 patients were categorised in cardiac.

14.0

SARAWAK

14.1 INTRODUCTION

Based on Department of Statistics Malaysia, the population in Sarawak is 2,907,500 people in 2020. The population group consists of Iban (29%), followed by Malay (23%), Chinese (22%), and Bidayuh (8%), Melanau (5%) and other Bumiputera (7%) and non-citizens with 6% of population. Sarawak's main centre of treatment is Hospital Umum Sarawak. There are five other districts hospitals in Sarawak namely Hospital Miri, Hospital Sibul, Hospital Bintulu, Hospital Lawas and Hospital Limbang. All district hospitals have their own specialist-in-charge, except for Hospital Lawas.

14.2 PATIENT DEMOGRAPHICS

There are 279 patients registered included 25 death cases in the MTR. 15 patients have received stem cell transplant. Most patients (64.96%) received treatment at Hospital Umum Sarawak. There are 16 new cases registered in 2020 that are six paediatric patients and 10 adult patients.

Table 14.1: Distribution of Patients in Sarawak by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Umum Sarawak	165	64.96
Hospital Miri	30	11.81
Hospital Lawas	18	7.09
Hospital Sibul	13	5.12
Hospital Limbang	12	4.72
Hospital Bintulu	12	4.72
Hospital Lundu	2	0.79
Hospital Saratok	1	0.39
Hospital Sarikei	1	0.39
Total	254	100.00

Table 14.2: Distribution of Patients in Sarawak by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	219
Cured By Stem Cell Therapy	15
Total	234
Lost To Follow Up	20
Total	254
Deaths in 2020	3
Cumulative Reported Deaths	25

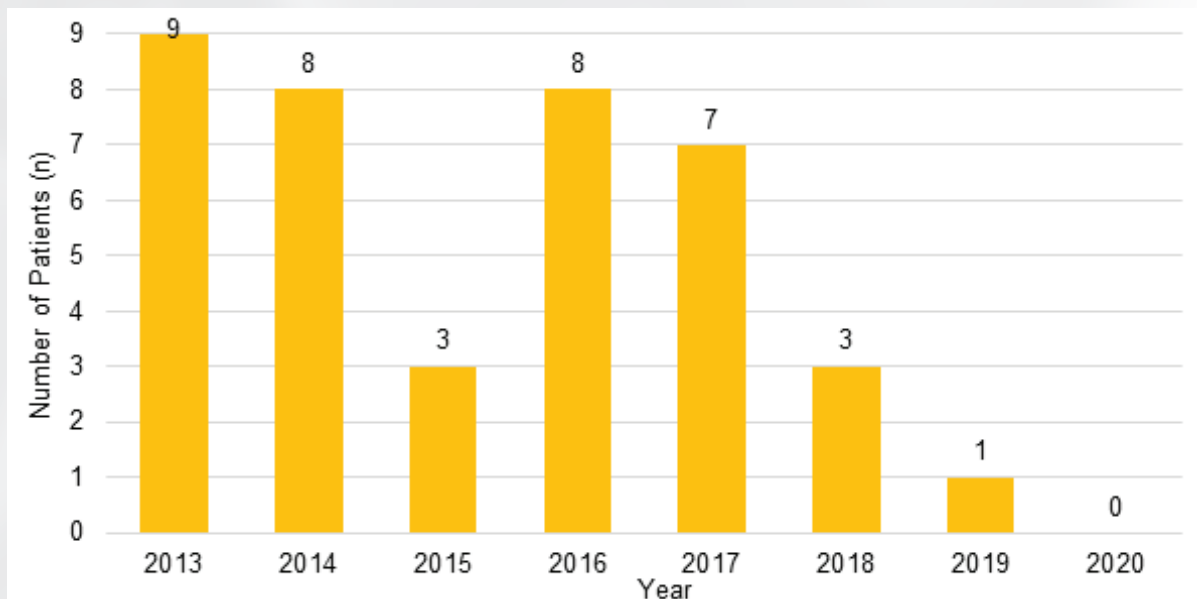


Figure 14.1: Distribution of Thalassaemia Births in Sarawak by Year

14.2.1 Age Groups

The youngest patient in Sarawak is 1 year old with β -thalassaemia major diagnosed at 9 months old and the oldest patient is 77 years old with Hb H disease. Patients below two years old are usually transfusion dependent. Most of the older patients in Sarawak have Hb H disease. They do not require any transfusion and some would require chelation at a later age when the serum ferritin level reaches above 800 ng/mL.

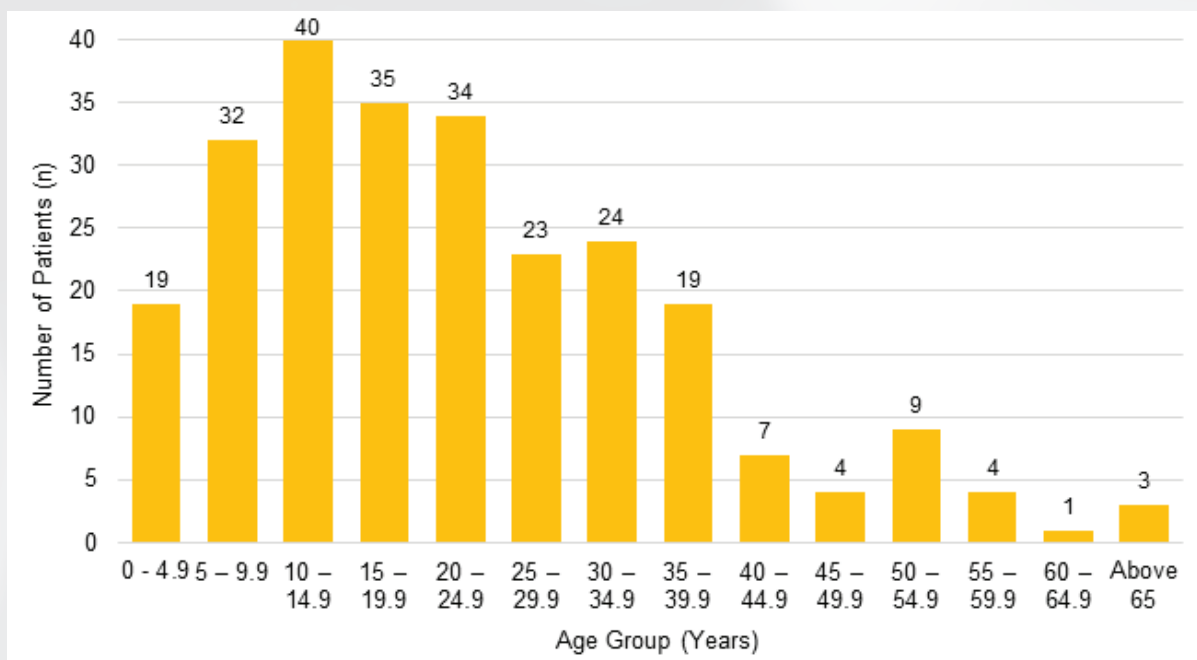


Figure 14.2: Distribution of Patients in Sarawak by Age Group

Table 14.3: 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	91	β -Thalassaemia Major	34	37.36
		β -Thalassaemia Intermedia	3	3.30
		HbE/ β -Thalassaemia	28	30.77
		Hb H Disease	26	28.57
		Others	0	0.00
15-29.9	92	β -Thalassaemia Major	44	47.83
		β -Thalassaemia Intermedia	7	7.61
		HbE/ β -Thalassaemia	19	20.65
		Hb H Disease	22	23.91
		Others	0	0.00
30-44.9	50	β -Thalassaemia Major	18	36.00
		β -Thalassaemia Intermedia	3	6.00
		HbE/ β -Thalassaemia	5	10.00
		Hb H Disease	24	48.00
		Others	0	0.00
45-59.9	17	β -Thalassaemia Major	0	0.00
		β -Thalassaemia Intermedia	2	11.76
		HbE/ β -Thalassaemia	3	17.65
		Hb H Disease	12	70.59
		Others	0	0.00
60 and above	4	β -Thalassaemia Major	0	0.00
		β -Thalassaemia Intermedia	0	0.00
		HbE/ β -Thalassaemia	0	0.00
		Hb H Disease	4	100.00
		Others	0	0.00
Total			254	

14.2.2 Gender

The gender distribution of thalassaemia patients in Sarawak consists of 122 males (48.03%) and 132 females (51.97%).

Table 14.4: Distribution of Patients in Sarawak According to Gender by Centre

Centre	Total Number of Patients	Male		Female	
		No.	%	No.	%
Hospital Umum Sarawak	165	70	27.56	95	37.40
Hospital Miri	30	18	7.09	12	4.72
Hospital Lawas	18	10	3.94	8	3.15
Hospital Sibul	13	5	1.97	8	3.15
Hospital Limbang	12	9	3.54	3	1.18
Hospital Bintulu	12	7	2.76	5	1.97
Hospital Lundu	2	2	0.79	0	0.00
Hospital Saratok	1	1	0.39	0	0.00
Hospital Sarikei	1	0	0.00	1	0.39
Total	254	122	48.03	132	51.97

14.2.3 Ethnic Group

The ethnic groups with the largest number of thalassaemia patients in Sarawak are Malay (47.64%) and Chinese (35.04%). It must be noted that the Iban do not carry the β -globin gene mutation, but instead carry the α -globin gene mutation. The majority of the Kedayan patients carry the Filipino β^0 -deletion mutation.

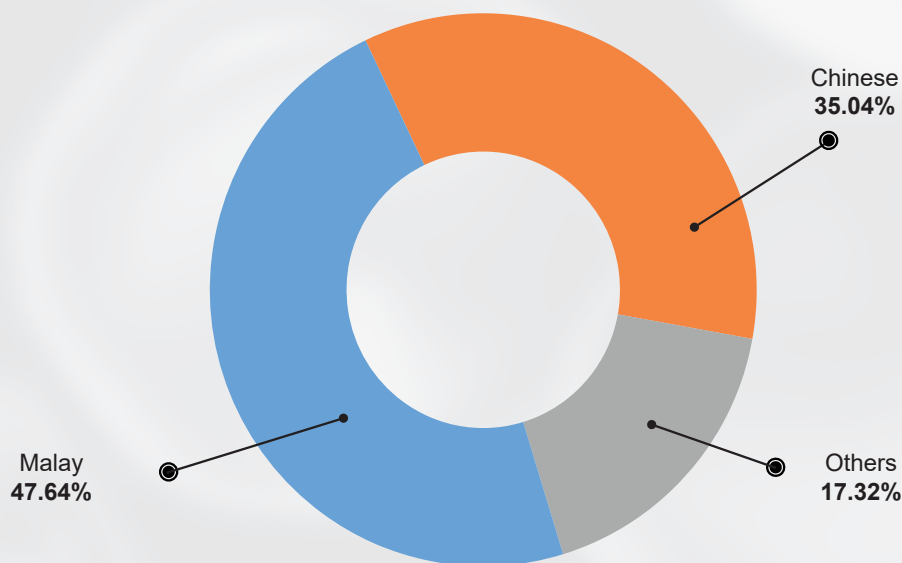


Figure 14.3: Distribution of Patients in Sarawak by Ethnic Group

Table 14.5: Distribution of Patients in Sarawak According to Ethnic Group by Centre

Centre	Total Number of Patients	Malay		Chinese		Others	
		No.	%	No.	%	No.	%
Hospita Umum Sarawak	165	83	32.68	64	25.20	18	7.09
Hospital Miri	30	8	3.15	9	3.54	13	5.12
Hospital Lawas	18	16	6.30	0	0.00	2	0.79
Hospital Sibul	13	0	0.00	12	4.72	1	0.39
Hospital Limbang	12	9	3.54	0	0.00	3	1.18
Hospital Bintulu	12	3	1.18	3	1.18	6	2.36
Hospital Lundu	2	2	0.79	0	0.00	0	0.00
Hospital Saratok	1	0	0.00	0	0.00	1	0.39
Hospital Sarikei	1	0	0.00	1	0.39	0	0.00
Total	254	121	47.64	89	35.04	44	17.32

14.3 DIAGNOSIS

The main diagnosis of thalassaemia patients in Sarawak is β -thalassaemia major, which make up of 37.80% patients followed by Hb H disease (34.65%), HbE/ β -thalassaemia (21.65%) and β -thalassaemia intermedia (5.91%). The prevalence of Hb H disease is not accurately reflected in this data, as no screening was performed and patients were incidentally noted to be pale. Most of the Hb H patients are asymptomatic.

No β -globin gene mutation was detected among the Iban population. Instead, the Iban harbours the β -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^0 -deletion mutation.

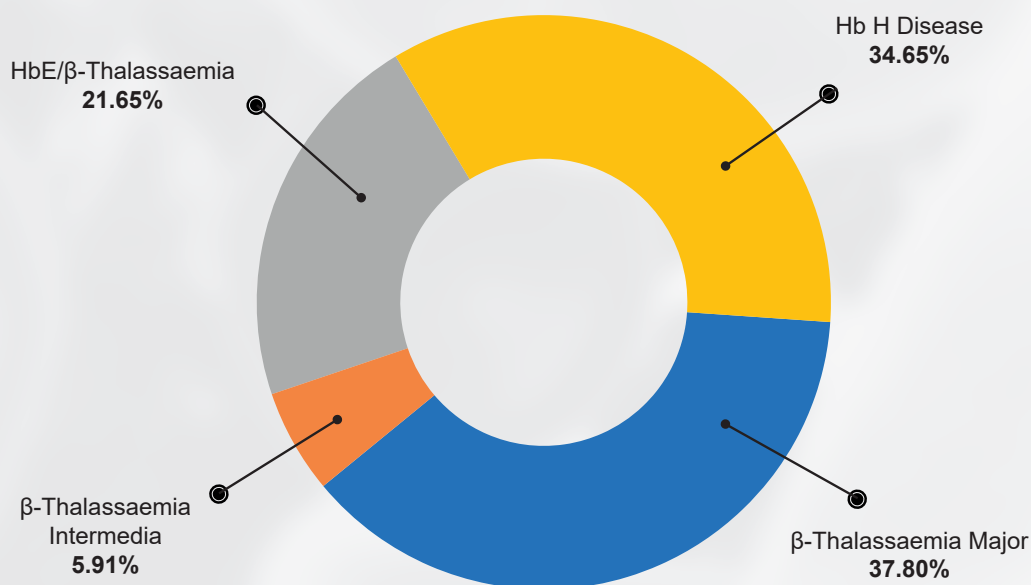


Figure 14.4: Distribution of Patients in Sarawak by Diagnosis

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

Centre	Total Number of Patients	β-Thalassaemia major		β-Thalassaemia intermedia		HbE/β-Thalassaemia		Hb H Disease	
		No.	%	No.	%	No.	%	No.	%
Hospita Umum Sarawak	165	44	17.32	8	3.15	44	17.32	69	27.17
Hospital Miri	30	21	8.27	1	0.39	2	0.79	6	2.36
Hospital Lawas	18	7	2.76	2	0.79	6	2.36	3	1.18
Hospital Sibul	13	10	3.94	2	0.79	0	0.00	1	0.39
Hospital Limbang	12	9	3.54	1	0.39	0	0.00	2	0.79
Hospital Bintulu	12	3	1.18	1	0.39	3	1.18	5	1.97
Hospital Lundu	2	2	0.79	0	0.00	0	0.00	0	0.00
Hospital Saratok	1	0	0.00	0	0.00	0	0.00	1	0.39
Hospital Sarikei	1	0	0.00	0	0.00	0	0.00	1	0.39
Total	254	96	37.80	15	5.91	55	21.65	88	34.65

Table 14.7: 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	96	Malay	30	11.81
		Chinese	51	20.08
		Iban	0	0.00
		Bidayuh	1	0.39
		Kedayan	11	4.33
		Pribumi Sarawak	2	0.79
		Kadazan Dusun	1	0.39
β-Thalassaemia Intermedia	15	Malay	7	2.76
		Chinese	6	2.36
		Iban	0	0.00
		Bidayuh	0	0.00
		Kedayan	1	0.39
		Pribumi Sarawak	0	0.00
		Pribumi Sabah	1	0.39
HbE/β-Thalassaemia	55	Malay	46	18.11
		Chinese	6	2.36
		Iban	1	0.39
		Bajau	1	0.39
		Bidayuh	1	0.39
Hb H Disease	88	Malay	38	14.96
		Chinese	26	10.24
		Iban	18	7.09
		Bidayuh	4	1.57
		Melanau	1	0.39
		Pribumi Sarawak	1	0.39
Total			254	100.00

14.4 TREATMENT

14.4.1 Iron Chelation Therapy

There are 125 patients in Sarawak who are receiving iron chelation therapy. Of these, 39.20% patients were prescribed DFX. All newly diagnosed patients in Sarawak were prescribed DFX monotherapy by two years of age if the serum ferritin level is higher than 1,000 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 23.20% patients were received a combination of DFO and DFP followed with 26.40% were prescribed with 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, 5.60% of the patients are prescribed with DFO monotherapy and 3.20% received a combination of DFO and DFX. Only three (2.40%) patients were on a combination of DFP and DFX.

Patients who received combination therapy had poor iron loading in the liver and cardiac or high serum ferritin levels, which were not responsive to chelation by monotherapy. Fifteen NTD patients in Sarawak required chelation. All of them are given DFP monotherapy due to high serum ferritin levels (more than 800 ng/mL). Ten of these patients required transfusions every 2-3 months. All 15 patients are aged above 20 years old.

Table 14.8: Distribution of Patients in Sarawak by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	7	5.60
DFP only	33	26.40
DFX only	49	39.20
DFO + DFP	29	23.20
DFP + DFX	3	2.40
DFO + DFX	4	3.20
DFO + DFP + DFX	0	0.00
Total	125	100.00

Table 14.9: 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	69	DFO only	4	3.20
		DFP only	21	16.80
		DFX only	24	19.20
		DFO + DFP	15	12.00
		DFP + DFX	1	0.80
		DFO + DFX	4	3.20
		DFO + DFP + DFX	0	0.00
Hospital Miri	16	DFO only	1	0.80
		DFP only	2	1.60
		DFX only	9	7.20
		DFO + DFP	4	3.20
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Lawas	12	DFO only	0	0.00
		DFP only	2	1.60
		DFX only	3	2.40
		DFO + DFP	7	5.60
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Limbang	9	DFO only	0	0.00
		DFP only	3	2.40
		DFX only	4	3.20
		DFO + DFP	1	0.80
		DFP + DFX	1	0.80
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Sibul	8	DFO only	1	0.80
		DFP only	1	0.80
		DFX only	5	4.00
		DFO + DFP	1	0.80
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Bintulu	8	DFO only	0	0.00
		DFP only	3	2.40
		DFX only	4	3.20
		DFO + DFP	0	0.00
		DFP + DFX	1	0.80
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Hospital Lundu	2	DFO only	1	0.80
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	0.80
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Sarikei	1	DFO only	0	0.00
		DFP only	1	0.80
		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			125	100.00

Table 14.10: 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	44	DFO Only	2	4.55
		DFP Only	4	9.09
		DFX Only	36	81.82
		DFO + DFP	1	2.27
		DFP + DFX	1	2.27
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	2.27
15-29.9	48	DFO Only	3	6.25
		DFP Only	11	22.92
		DFX Only	9	18.75
		DFO + DFP	22	45.83
		DFP + DFX	0	0.00
		DFO + DFX	3	6.25
		DFO + DFP + DFX	0	0.00
30-44.9	20	DFO Only	2	10.00
		DFP Only	6	30.00
		DFX Only	4	20.00
		DFO + DFP	6	30.00
		DFP + DFX	2	10.00

		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-59.9	10	DFO Only	9	90.00
		DFP Only	0	0.00
		DFX Only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	10.00
		DFO + DFP + DFX	0	0.00
		60 and above	3	DFO Only
DFP Only	3			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			125	

14.4.2 Serum Ferritin Level

A total of 187 patients in Sarawak had their serum ferritin level recorded in 2020. Of these, 116 (62.03%) are TDT patients and 71 (37.97%) are NTD patients. A total of 81 (43.32%) patients has a serum ferritin level lower than 1,000 ng/mL, 68 patients (36.36%) have a serum ferritin level between 1,000-2,499 ng/mL, and 19 (10.16%) patients have serum ferritin level between 2,500-4,999 ng/mL. Besides that, 16 (8.56%) patients have a serum ferritin level between 5,000-9,999 ng/mL and three (1.60%) patients with serum ferritin above 10,000 ng/mL.

Most patients (149 out of 187 patients, 79.68%) from regional and tertiary centres have a serum ferritin level lower than 2,500 ng/mL, whereas 38 out of 161 patients (20.32%) have a serum ferritin level above 2,500 ng/mL. The patients in Hospital Umum Sarawak and Hospital Sibul 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. There are 79 TDT patients in Sarawak have a serum ferritin level < 2,500 ng/mL. Two patients in Hospital Umum Sarawak and one patient in Hospital Sibul have serum ferritin level > 10,000 ng/mL.

Table 14.11: Distribution of TDT and NTD T 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	110	56	29.95	38	20.32	7	3.74	7	3.74	2	1.07
Hospital Miri	26	13	6.95	7	3.74	5	2.67	1	0.53	0	0.00
Hospital Sibul	10	1	0.53	7	3.74	1	0.53	0	0.00	1	0.53
Hospital Bintulu	10	2	1.07	7	3.74	0	0.00	1	0.53	0	0.00
Hospital Lawas	16	5	2.67	2	1.07	5	2.67	4	2.14	0	0.00
Hospital Limbang	11	3	1.60	5	2.67	1	0.53	2	1.07	0	0.00
Hospital Lundu	2	0	0.00	1	0.53	0	0.00	1	0.53	0	0.00
Hospital Saratok	1	1	0.53	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Sarikei	1	0	0.00	1	0.53	0	0.00	0	0.00	0	0.00
Total	187	81	43.32	68	36.36	19	10.16	16	8.56	3	1.60

14.5 COMPLICATIONS AND DEATHS

14.5.1 Complications

There are 67 TDT and 96 NTDT patients above 15 years old in Sarawak.

Based on Table 14.12, two adult patients were diagnosed with hepatitis B, and another 10 patients were diagnosed with hepatitis C in 2019. It is unknown whether the transmissions were due to previous transfusions, or due to high-risk behaviors.

Table 14.12: Distribution of Patients in Sarawak According to Transfusion Transmissible Infections

Complications	2007-2019	2020
Hepatitis C: Anti HCV	10	0
HCV RNA	0	0
Hepatitis B	2	0
HIV	0	0
Total	12	0

Based on Table 14.13, 27 were diagnosed with delayed puberty and were put on hormonal replacement therapy. These patients have been poorly chelated in the past. A total of 54 patients were noted to have short stature, but further information is acquired to determine whether this is secondary to growth hormone deficiency or familial short stature.

Table 14.13: Distribution of Patient in Sarawak According to Endocrine Complications

Complications	2007-2019
Short Stature	54
Delayed Puberty	27
Hypothyroid	11
Diabetes Mellitus	6
Total	98

14.5.2 Iron Deposition in Heart and Liver Complications

Based on Table 14.14, there are 63 patients had cardiac MRI T2* performed. 51 (80.95%) patients have a normal iron loading of cardiac. Some of these patients had shown an initial result of less than 20 ms, but has since improved over the years with intensive chelation. In 2020, 17 patients had MRI T2* scans performed. Two of these patients had result less than 10 ms. These two patients were known to be poorly compliant to chelation therapy for many years. An additional two patients had an MRI T2* of between 10-20 ms. Most of these patients have endocrine problems. Based on Table 14.15, 62 patients had liver MRI performed.

Table 14.14: Distribution of Patients in Sarawak According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Umum Sarawak	43	36	57.14	2	3.17	0	0.00	5	7.94
Hospital Miri	8	7	11.11	1	1.59	0	0.00	0	0.00
Hospital Sibul	7	6	9.52	0	0.00	0	0.00	1	1.59
Hospital Bintulu	2	1	1.59	1	1.59	0	0.00	0	0.00
Hospital Lundu	2	0	0.00	2	3.17	0	0.00	0	0.00
Hospital Limbang	1	1	1.59	0	0.00	0	0.00	0	0.00
Hospital Lawas	0	0	0.00	0	0.00	0	0.00	0	0.00
Total	63	51	80.95	6	9.52	0	0.00	6	9.52

Table 14.15: Distribution of Patients in Sarawak According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Umum Sarawak	43	2	3.23	16	25.81	21	33.87	4	6.45
Hospital Miri	8	2	3.23	3	4.84	2	3.23	1	1.61
Hospital Sibul	7	1	1.61	1	1.61	4	6.45	1	1.61
Hospital Lundu	2	0	0.00	0	0.00	0	0.00	2	3.23
Hospital Bintulu	1	0	0.00	0	0.00	1	1.61	0	0.00
Hospital Limbang	1	0	0.00	1	1.61	0	0.00	0	0.00
Hospital Lawas	0	0	0.00	0	0.00	0	0.00	0	0.00
Total	62	5	8.06	21	33.87	28	45.16	8	12.90

14.5.3 Death Cases

A total of 25 patients had died in Sarawak. One patient died in December 2019 and three patients were reported dead in 2020.

Table 14.16: Cumulative Known Causes of Death in Sarawak

Causes of Death	Number of Patients (n)
Cardiac	17
Infection	3
Thalassaemia	1
Malignancy	1
Thrombosis	1
Liver Disease	1
Others	1
Total	25

14.6 CONCLUSION

Based on the data collection, there was a 4.1% increment in the number of new cases registered in the MTR in 2020. Of the 16 new cases registered, six are children (two were diagnosed with Hb H disease, one with β -thalassaemia major, and three 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 20-24.9 years old, which differs from the median age for last year's as there are still new cases registered for the paediatric and adult cohort each year.

Most patients (80%) in Sarawak have a serum ferritin level < 2,500 ng/mL. In the TDT cohorts, 68% of patients have a serum ferritin of < 2,500 ng/mL, of which 20% have a serum ferritin level < 1,000 ng/ml. A majority of patients with elevated serum ferritin level (> 2,500 ng/mL) are adult patients above 15 years old, who are in 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 such as 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. On the other hand, Lawas consistently recorded the highest number of patients with serum ferritin level > 2,500 ng/mL.

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. A total of 67 TDT patients are above 15 years old. Of these, approximately 50% patients with delayed puberty and short stature. Better chelation during childhood is expected to reduce endocrine complications over the years.

There are 63 patients who had cardiac MRI T2* scan performed. 19% have an MRI T2* result below 20 ms. Some patients had an MRI T2* results of < 20 ms, but was reversed over the years with intensive chelation. Since the beginning of the registry, Sarawak has recorded 25 deaths, mostly due to cardiac complications. Of note, there was a mortality attributed to severe haemolytic anemia 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.



15.0
SELANGOR

15.1 INTRODUCTION

Selangor is the eighth largest states in Peninsular Malaysia with an estimated population of 6,538,000 in 2020 based on 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.

15.2 PATIENT DEMOGRAPHICS

Table 15.1 shows that there are 1,383 living thalassaemia patients and they received transfusions and follow-up care in the hospitals listed.

Table 15.1: Distribution of Patients in Selangor by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Ampang	761	55.03
Hospital Tengku Ampuan Rahimah	295	21.33
Hospital Selayang	100	7.23
Hospital Sungai Buloh	78	5.64
Hospital Kajang	56	4.05
Hospital Serdang	43	3.11
Hospital Shah Alam	22	1.59
Hospital Banting	17	1.23
Hospital Tengku Ampuan Jemaah	6	0.43
Hospital Kuala Kubu Bharu	3	0.22
Hospital Tanjung Karang	2	0.14
Total	1383	100.00

Table 15.2 shows the distribution of patients based on vital status. There are a total of 1,123 alive patients, 11 patients with haemopoietic stem cell transplant and 249 patients who lost to follow up.

Table 15.2: Distribution of Patients in Selangor by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	1123
Cured by Stem Cell Therapy	11
Total	1134
Lost to Follow Up	249
Total	1383
Death in 2020	7
Cumulative Reported Deaths	89

Figure 15.1 showed a decreasing trend from 2013 to 2020. The decreased number is reflected by increase awareness of thalassaemia and its prevention among the families and the public.

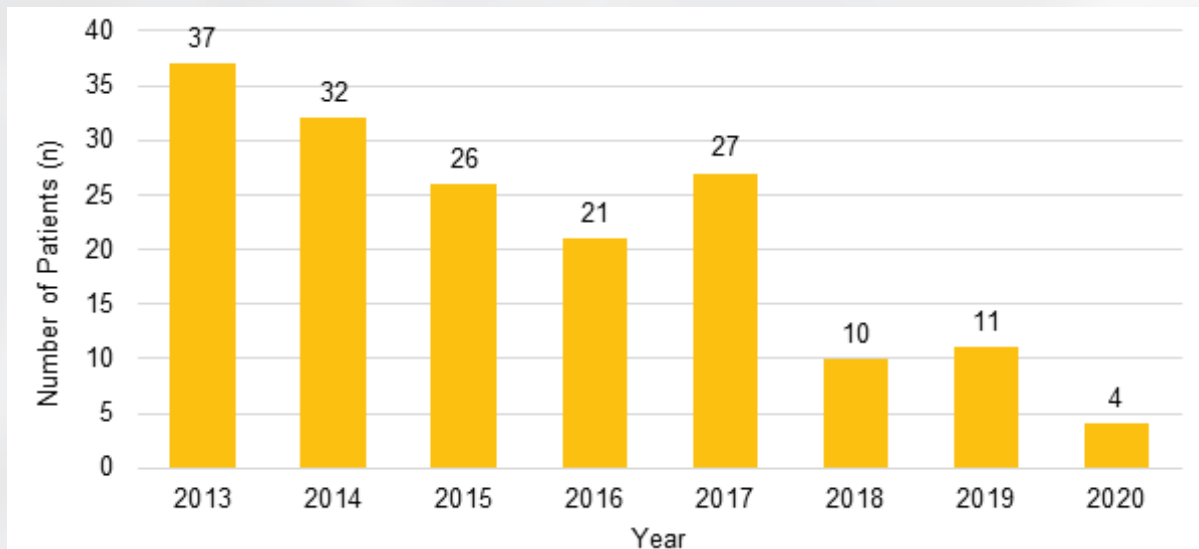


Figure 15.1: Distribution of Thalassaemia Births in Selangor by Year

15.2.1 Age Groups

Approximately 33.41% of thalassaemia patients in Selangor are below 20 years old, and thus classified under the paediatric and adolescent age categories. About 66.59% of the total thalassaemia cases in Selangor consist of adult patients above 21 years old. The oldest patient in Selangor is an 89 years old Malay gentleman diagnosed with Hb H disease. Meanwhile, the youngest patient is a five months old Malay girl diagnosed with Hb H disease and followed up at Hospital Sungai Buloh.

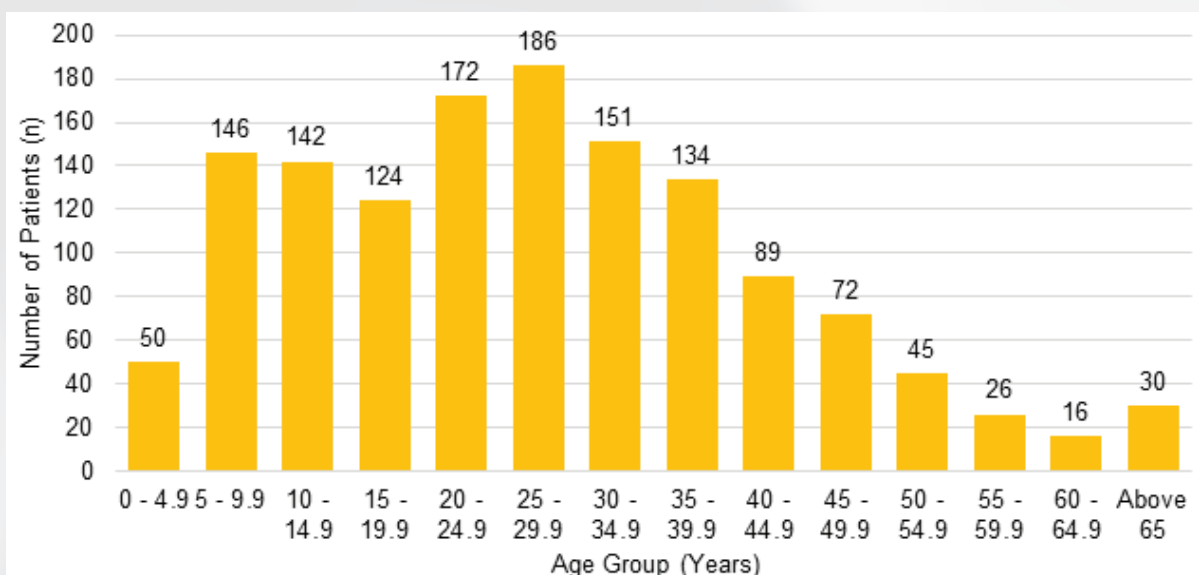


Figure 15.2: Distribution of Patients in Selangor by Age Group

Table 15.3: Distribution of Patients in Selangor According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patients (n)	Percentage (%)
0-14.9	338	β-Thalassaemia Major	60	17.75
		β-Thalassaemia Intermedia	19	5.62
		HbE/β-Thalassaemia	166	49.11
		Hb H Disease	70	20.71
		Others	23	6.80
15-29.9	482	β-Thalassaemia Major	108	22.41
		β-Thalassaemia Intermedia	31	6.43
		HbE/β-Thalassaemia	220	45.64
		Hb H Disease	113	23.44
		Others	10	2.07
30-44.9	374	β-Thalassaemia Major	56	14.97
		β-Thalassaemia Intermedia	25	6.68
		HbE/β-Thalassaemia	164	43.85
		Hb H Disease	123	32.89
		Others	6	1.60
45-59.9	143	β-Thalassaemia Major	7	4.90
		β-Thalassaemia Intermedia	17	11.89
		HbE/β-Thalassaemia	52	36.36
		Hb H Disease	65	45.45
		Others	2	1.40
60 and above	46	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	6	13.04
		HbE/β-Thalassaemia	10	21.74
		Hb H Disease	29	63.04
		Others	1	2.17
Total			1383	

15.2.2 Gender

Based on Table 15.4, there are slightly more female than male thalassaemia patients in Selangor. There are 758 females (54.81%) and 625 males (45.19%), with a female to male ratio of 1.2:1.0.

Table 15.4: Distribution of Patients in Selangor According to Gender by Centre

Centre	Total Number of Patients	Male		Female	
		No.	%	No.	%
Hospital Ampang	761	305	22.05	456	32.97
Hospital Tengku Ampuan Rahimah	295	134	9.69	161	11.64
Hospital Selayang	100	53	3.83	47	3.40
Hospital Sungai Buloh	78	51	3.69	27	1.95
Hospital Kajang	56	30	2.17	26	1.88
Hospital Serdang	43	23	1.66	20	1.45
Hospital Shah Alam	22	11	0.80	11	0.80
Hospital Banting	17	13	0.94	4	0.29
Hospital Tengku Ampuan Jemaah	6	3	0.22	3	0.22
Hospital Kuala Kubu Bharu	3	2	0.14	1	0.07
Hospital Tanjung Karang	2	0	0.00	2	0.14
Total	1383	625	45.19	758	54.81

15.2.3 Ethnic Group

Based on Figure 15.3, Malay forms the largest group of patients in Selangor with 1,090 patients (78.81%), followed by the Chinese with 201 patients (14.53%), Indian with eight patients (0.58%), Kadazan Dusun with 20 patients (1.45%), and Orang Asli with seven patients (0.51%). Other ethnic groups consist of 57 patients (4.12%). Those categorised under “Others” included Melanau, Murut, Bajau, Bidayuh, Thais, mixed, foreigners, Rungus, Pribumi Sabah and others.

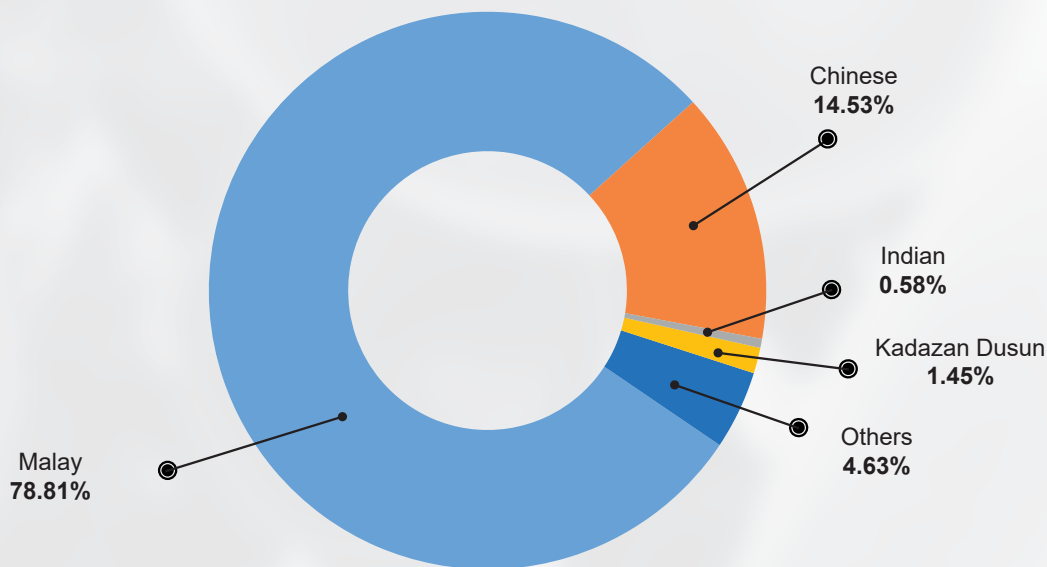


Figure 15.3: Distribution of Patients in Selangor by Ethnic Group

15.3 DIAGNOSIS

The majority of patients have HbE/ β -thalassaemia (612 patients, 44.25%) in Selangor. This is followed by Hb H disease (400 patients, 28.92%), β -thalassaemia major (231 patients, 16.70%), and β -thalassaemia intermedia (98 patients, 7.09%). The remaining 42 patients (3.04%) are being followed up for other forms of haemoglobinopathy. This is depicted in Figure 15.4.

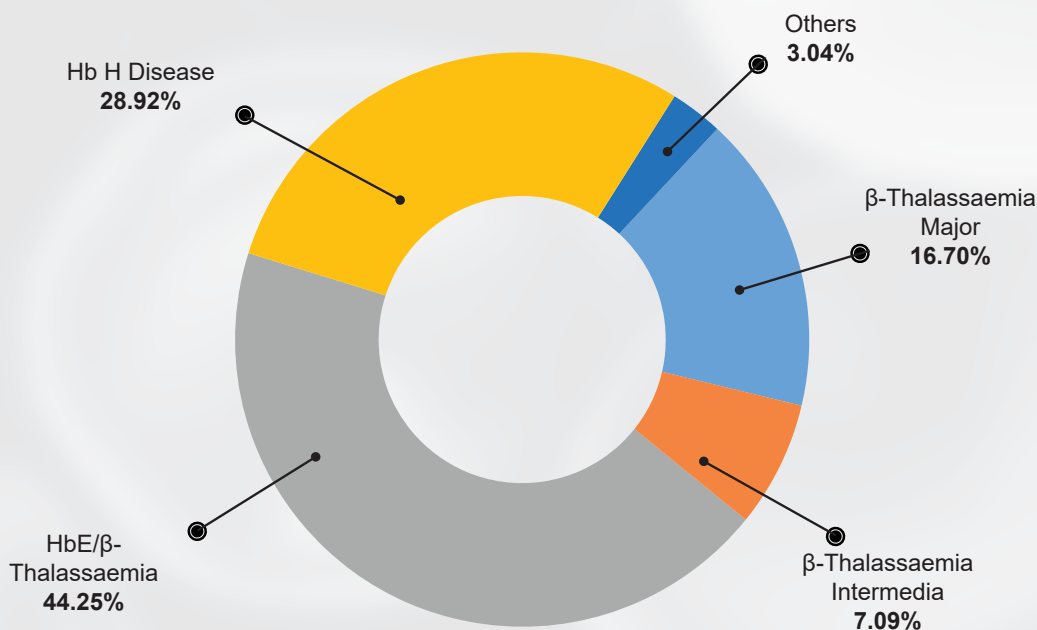


Figure 15.4: Distribution of Patients in Selangor by Diagnosis

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

Centre	Total Number of Patients	β-Thalassaemia Major		β-Thalassaemia Intermedia		HbE/β-Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Ampang	761	132	9.54	43	3.11	318	22.99	258	18.66	10	0.72
Hospital Tengku Ampuan Rahimah	295	42	3.04	36	2.60	143	10.34	63	4.56	11	0.80
Hospital Selayang	100	16	1.16	6	0.43	31	2.24	37	2.68	10	0.72
Hospital Sungai Buloh	78	16	1.16	3	0.22	41	2.96	13	0.94	5	0.36
Hospital Kajang	56	11	0.80	4	0.29	22	1.59	17	1.23	2	0.14
Hospital Serdang	43	8	0.58	5	0.36	23	1.66	4	0.29	3	0.22
Hospital Shah Alam	22	2	0.14	1	0.07	14	1.01	4	0.29	1	0.07
Hospital Banting	17	3	0.22	0	0.00	11	0.80	3	0.22	0	0.00
Hospital Tengku Ampuan Jemaah	6	0	0.00	0	0.00	6	0.43	0	0.00	0	0.00
Hospital Kuala Kubu Bharu	3	1	0.07	0	0.00	1	0.07	1	0.07	0	0.00
Hospital Tanjung Karang	2	0	0.00	0	0.00	2	0.14	0	0.00	0	0.00
Total	1383	231	16.70	98	7.09	612	44.25	400	28.92	42	3.04

Table 15.6 shows that the majority of Malay patients have HbE/ β -thalassaemia with 556 patients (51.28%). Meanwhile, Chinese patients are more commonly diagnosed with Hb H disease by 91 patients (45.27%). Indian patients, the prominent diagnosis is under “Others” with only three patients (37.5%) which mostly is Hb Lapore Hollandia.

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	231	Malay	127	9.18
		Chinese	72	5.21
		Indian	2	0.14
		Kadazan Dusun	13	0.94
		Others	17	1.23
β -Thalassaemia Intermedia	98	Malay	79	5.71
		Chinese	12	0.87
		Indian	1	0.07
		Kadazan Dusun	2	0.14
		Others	4	0.29
HbE/ β -Thalassaemia	612	Malay	559	40.42
		Chinese	21	1.52
		Indian	1	0.07
		Kadazan Dusun	3	0.22
		Others	28	2.02
Hb H Disease	400	Malay	294	21.26
		Chinese	91	6.58
		Indian	1	0.07
		Kadazan Dusun	1	0.07
		Others	13	0.94
Others	42	Malay	31	2.24
		Chinese	5	0.36
		Indian	3	0.22
		Kadazan Dusun	1	0.07
		Others	2	0.14
Total			1383	100.00

15.4 TREATMENT

15.4.1 Iron Chelation Therapy

Table 15.7 shows a total of 886 thalassaemia patients who received iron chelation therapy in Selangor. The most common iron chelation therapy used is DFP monotherapy with 40.97% patients on this regime. The second most common regime is DFO + DFP combination therapy with 24.83% of patients on this. Combination therapy of DFP + DFX and DFO + DFX showed a percentage value of 2.6% and 2.03%, respectively. Only 0.45% patients receive a combination of all chelators. There are 497 patients who are not on iron chelation therapy.

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

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	75	8.47
DFP only	363	40.97
DFX only	183	20.65
DFO + DFP	220	24.83
DFP + DFX	23	2.60
DFO + DFX	18	2.03
DFO + DFP + DFX	4	0.45
Total	886	100.00

As shown in Table 15.8, the majority of patients who received iron chelator are 501 patients (56.52%) in Hospital Ampang, followed by 193 patients (21.78%) in Hospital Tuanku Ampuan Rahimah. Patients in Hospital Ampang also frequently received DFP and combination of DFP and DFO while DFP and DFX were frequently received by patients in HTAR, Klang.

DFX monotherapy is the commonest iron chelator used in Hospital Serdang, Hospital Selayang, Hospital Banting and Hospital Shah Alam as thalassaemia patients in these hospitals are of the paediatrics age group. Hospital Sungai Buloh and Hospital Kajang treat both pediatric and adolescent thalassaemia patients and they frequently use a combination of DFP + DFO therapy.

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	501	DFO only	33	3.72
		DFP only	262	29.57
		DFX only	33	3.72
		DFO + DFP	159	17.95
		DFP + DFX	6	0.68
		DFO + DFX	8	0.90
		DFO + DFP + DFX	0	0.00
Hospital Tengku Ampuan Rahimah	193	DFO only	6	0.68
		DFP only	72	8.13
		DFX only	64	7.22
		DFO + DFP	26	2.93
		DFP + DFX	16	1.81
		DFO + DFX	5	0.56
		DFO + DFP + DFX	4	0.45
Hospital Sungai Buloh	48	DFO only	3	0.34
		DFP only	14	1.58
		DFX only	10	1.13
		DFO + DFP	20	2.26
		DFP + DFX	0	0.00
		DFO + DFX	1	0.11
		DFO + DFP + DFX	0	0.00
Hospital Selayang	42	DFO only	3	0.34
		DFP only	5	0.56
		DFX only	30	3.39
		DFO + DFP	1	0.11
		DFP + DFX	1	0.11
		DFO + DFX	2	0.23
		DFO + DFP + DFX	0	0.00
Hospital Serdang	39	DFO only	18	2.03
		DFP only	1	0.11
		DFX only	19	2.14
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.11
		DFO + DFP + DFX	0	0.00
Hospital Kajang	27	DFO only	6	0.68
		DFP only	3	0.34
		DFX only	6	0.68
		DFO + DFP	11	1.24
		DFP + DFX	0	0.00
		DFO + DFX	1	0.11
		DFO + DFP + DFX	0	0.00

Hospital Banting	16	DFO only	3	0.34
		DFP only	4	0.45
		DFX only	7	0.79
		DFO + DFP	2	0.23
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Shah Alam	13	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	13	1.47
		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	4	DFO only	2	0.23
		DFP only	1	0.11
		DFX only	1	0.11
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kuala Kubu Bharu	3	DFO only	1	0.11
		DFP only	1	0.11
		DFX only	0	0.00
		DFO + DFP	1	0.11
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		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
Total			886	100.00

Table 15.9 shows patients up to 70 years old with iron chelation therapy and the highest number of patients that received the treatment is at 15-29.9 years old age group (36.91%). The thalassaemia patients between 0 and 15 years mostly received DFX (15.12%). Meanwhile, patients between 15 and 70 years commonly received DFP and combination of DFP + DFO. Paediatric patients who previously were on DFX are usually switched to DFO or DFP upon transfer to adult physician care as there is no additional funding to support DFX in this cohort of patients.

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	224	DFO only	30	13.39
		DFP only	22	9.82
		DFX only	134	59.82
		DFO + DFP	20	8.93
		DFP + DFX	12	5.36
		DFO + DFX	5	2.23
		DFO + DFP + DFX	1	0.45
15-29.9	327	DFO only	22	6.73
		DFP only	131	40.06
		DFX only	36	11.01
		DFO + DFP	118	36.09
		DFP + DFX	9	2.75
		DFO + DFX	8	2.45
		DFO + DFP + DFX	3	0.92
30-44.9	234	DFO only	17	7.26
		DFP only	140	59.83
		DFX only	4	1.71
		DFO + DFP	69	29.49
		DFP + DFX	1	0.43
		DFO + DFX	3	1.28
		DFO + DFP + DFX	0	0.00

45-59.9	79	DFO only	6	7.59
		DFP only	53	67.09
		DFX only	5	6.33
		DFO + DFP	12	15.19
		DFP + DFX	1	1.27
		DFO + DFX	2	2.53
		DFO + DFP + DFX	0	0.00
60 and above	22	DFO only	0	0.00
		DFP only	17	77.27
		DFX only	4	18.18
		DFO + DFP	1	4.55
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			886	

15.4.2 Serum Ferritin Level

Table 15.10 shows a total of 597 TDT patients who had their serum ferritin levels measured in 2020. Hospital Ampang and Hospital Tengku Ampuan Rahimah are centres that have the highest number of TDT patients in Selangor. About 330 of TDT patients (55.27%) had serum ferritin level below 2,500 ng/mL, and 267 TDT patients (44.72%) had serum ferritin levels more than 2,500 ng/mL. These results showed that TDT patients in Selangor have adequate chelation therapy. It is recommended to maintain serum ferritin level lower than 2,500 ng/mL to prevent iron induced-organ injury.

Table 15.10: Distribution of TDT 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	277	52	8.71	90	15.08	66	11.06	55	9.21	14	2.35
Hospital Tengku Ampuan Rahimah	145	22	3.69	61	10.22	42	7.04	18	3.02	2	0.34
Hospital Sungai Buloh	48	12	2.01	25	4.19	8	1.34	3	0.50	0	0.00
Hospital Serdang	39	2	0.34	12	2.01	19	3.18	5	0.84	1	0.17
Hospital Selayang	30	1	0.17	21	3.52	8	1.34	0	0.00	0	0.00
Hospital Kajang	25	3	0.50	5	0.84	10	1.68	5	0.84	2	0.34
Hospital Banting	16	2	0.34	8	1.34	4	0.67	2	0.34	0	0.00
Hospital Shah Alam	13	1	0.17	10	1.68	2	0.34	0	0.00	0	0.00
Hospital Tengku Ampuan Jemaah	3	0	0.00	2	0.34	1	0.17	0	0.00	0	0.00
Hospital Kuala Kubu Baru	1	0	0.00	1	0.17	0	0.00	0	0.00	0	0.00
Total	597	95	15.91	235	39.36	160	26.80	88	14.74	19	3.18

15.5 COMPLICATIONS AND DEATHS

15.5.1 Complications

Table 15.11 shows the transfusion transmissible infections of thalassaemia patients receiving treatment. There are 28 patients who have hepatitis C including three patients with recent infection, nine patients contracted hepatitis B and one patient with HIV.

Table 15.11: Distribution of Patients in Selangor According to Transfusion Transmissible Infections

Infections	2007-2019	2020
Hepatitis C: Anti HCV	25	0
HCV RNA	0	3
Hepatitis B	9	0
HIV	1	0
Total	35	3

Based on Table 15.12, 97 patients have short stature as a complication of chronic iron overload, 49 patients with hypothyroid and 42 patients have delayed puberty.

Table 15.12: Distribution of Patient in Selangor According to Endocrine Complication

Complications	Number of Patients (n)
Short Stature	97
Hypothyroid	49
Delayed Puberty	42
Diabetes Mellitus	34
Total	222

15.5.2 Iron Deposition in Heart and Liver Complications

Table 15.13 shows the liver and cardiac MRI T2* results of thalassaemia patients in Selangor. There are 84.11% patients with normal iron loading and only 7.95% have severe iron loading of the cardiac. Table 15.14 shows 47.52% patients with severe iron loading and only 6.79% with normal iron loading of the liver.

Table 15.13: Distribution of Patients in Selangor According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Ampang	402	331	54.80	16	2.65	19	3.15	36	5.96
Hospital Tengku Ampuan Rahimah	131	114	18.87	3	0.50	4	0.66	10	1.66
Hospital Selayang	22	21	3.48	1	0.17	0	0.00	0	0.00
Hospital Kajang	20	16	2.65	2	0.33	1	0.17	1	0.17
Hospital Serdang	16	14	2.32	0	0.00	1	0.17	1	0.17
Hospital Sungai Buloh	9	9	1.49	0	0.00	0	0.00	0	0.00
Hospital Tengku Ampuan Jemaah	2	1	0.17	1	0.17	0	0.00	0	0.00
Hospital Banting	2	2	0.33	0	0.00	0	0.00	0	0.00
Total	604	508	84.11	23	3.81	25	4.14	48	7.95

Table 15.14: Distribution of Patients in Selangor According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild /Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Ampang	402	32	5.30	74	12.25	91	15.07	205	33.94
Hospital Klang	131	5	0.83	22	3.64	44	7.28	60	9.93
Hospital Selayang	22	0	0.00	6	0.99	7	1.16	9	1.49
Hospital Kajang	20	2	0.33	6	0.99	6	0.99	6	0.99
Hospital Serdang	16	1	0.17	6	0.99	6	0.99	3	0.50
Hospital Sungai Buloh	9	0	0.00	2	0.33	3	0.50	4	0.66
Hospital Tengku Ampuan Jemaah	2	0	0.00	0	0.00	2	0.33	0	0.00
Hospital Banting	2	1	0.17	1	0.17	0	0.00	0	0.00
Total	604	41	6.78	117	19.37	159	26.32	287	47.52

15.5.3 Death Cases

Based on Table 15.15, there are 89 cumulative total of deaths of thalassaemia patients which includes five unknown causes of death due to unavailable data. There are seven patients who died in 2020. Four of the patients died due to cardiac, one with liver disease, followed by another with infections and one with endocrine complication.

Table 15.15: Cumulative Known Causes of Death in Selangor

Causes of Death	Number of Patients (n)
Cardiac	35
Infections	29
Liver Disease	8
Motor Vehicle Accident (MVA)	6
Malignancy	4
Endocrine Complications	2
Total	84

15.6 CONCLUSION

Hospital Ampang and Hospital Tengku Ampuan Rahimah are the two main centres with the highest number of thalassaemia patients registered. Based on ethnicity, Malay has the highest number of patients diagnosed with HbE/β-Thalassaemia followed by Chinese and Indian. Other ethnic groups treated in Selangor include Kadazan Dusun, Orang Asli, Melanau, Murut, Bajau, Bidayuh, Thais, mixed, foreigners, Rungus, Pribumi Sabah and others. Male and female are equally affected by thalassaemia and the highest number of patients is in the age group of 15-29.9 years old.

DFP is the most common iron chelation therapy used especially in adult centres and DFX is more frequently used in paediatric centres, namely, Hospital Selayang, Banting and Serdang. Sometimes the iron chelation therapy received by thalassaemia patients was given based on the additional budget. Over half of the TDT patients in Selangor receive adequate iron chelation therapy based on serum ferritin levels findings. More than 50% TDT patients show serum ferritin levels lower than 2,500 ng/mL which indicates adequate and compliance to chelation therapy.

The outcome of liver and cardiac MRI T2* scan shows that approximately 7.95% of the TDT patients with normal iron loading of cardiac and 47.52% of the patients with severe iron loading of liver. The highest number of death cases was 32 patients due to cardiac as a result of chronic iron overload. Endocrine complication was the highest complication followed by infections related.



16.0
TERENGGANU

16.1 INTRODUCTION

Based on Department of Statistics Malaysia website the population, in Terengganu by 2020 is 1.25 million. The state of Terengganu has eight districts, namely, Besut, Setiu, Dungun, Hulu Terengganu, Kuala Nerus, Marang, Kuala Terengganu and Kemaman. Hospital Sultanah Nur Zahirah, Kuala Terengganu, being the state hospital and the only tertiary hospital in Terengganu, receives the largest number of thalassaemia patients. Other hospitals in the state that offer thalassaemia care are hospital Dungun, Besut, Setiu, Kemaman and Hulu Terengganu.

Hospital Sultanah Nur Zahirah, Hospital Kemaman and Hospital Hulu Terengganu provides blood transfusion service in day care centres. Although the other district hospitals, such as 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 whereby transfusion is done in the ward but will be discharged on the same day.

16.2 PATIENT DEMOGRAPHICS

Data on vital status of patient are consists of status active treatment, lost to follow-up or cured by transplant. For 2020, the total number of thalassaemia patients in Terengganu is 364 patients. TDT patients are 249 and 115 patients are non-transfusion dependent (NTDT). Table 16.1 summarised the number of patients by centre and Table 16.2 summarised 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	223	61.26
Hospital Hulu Terengganu	37	10.16
Hospital Kemaman	35	9.62
Hospital Besut	27	7.42
Hospital Dungun	24	6.59
Hospital Setiu	18	4.95
Total	364	100

Table 16.2: Distribution of Patients in Terengganu by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	321
Cured by Stem Cell Therapy	3
Total	324
Lost to Follow Up	40
Total	364
Death in 2020	1
Cumulative Reported Deaths	30

Up until November 2020, only one death of thalassaemia patient was recorded. Three patients underwent haematopoietic stem cell transplantation (HSCT) between 2017 and 2019. There were six new cases diagnosed in 2020 and all of them are from HSNZ, Kuala Terengganu.

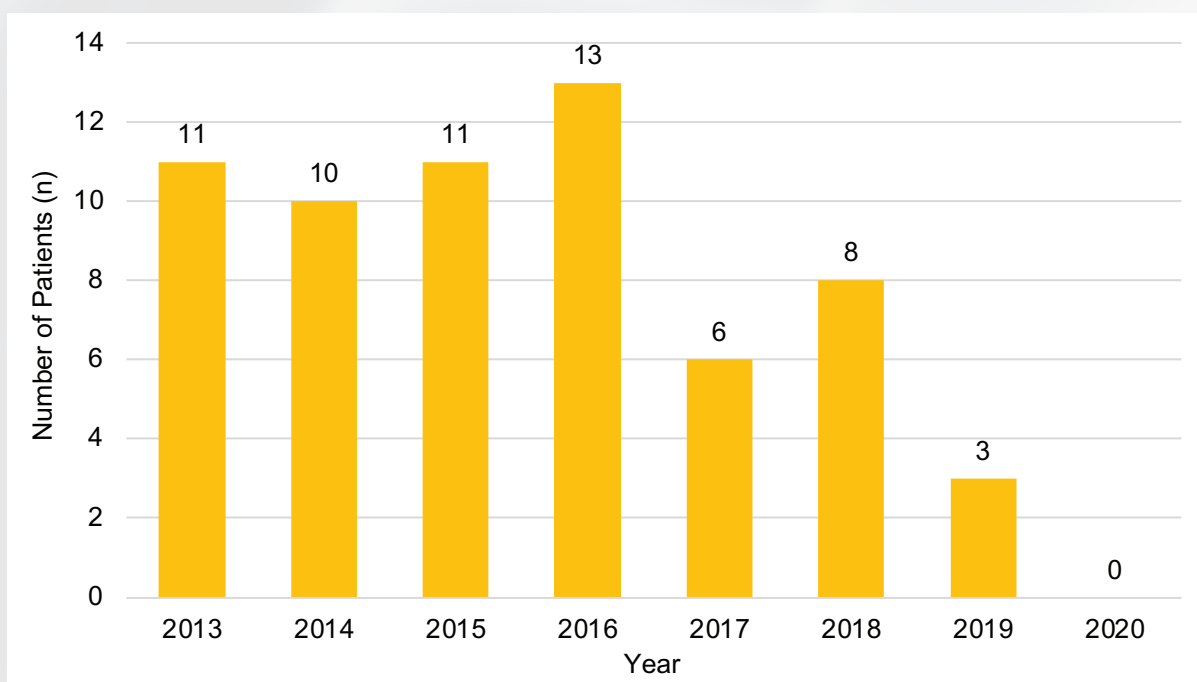


Figure 16.1: Distribution of Thalassaemia Births in Terengganu by Year

16.2.1 Age Groups

The majority of thalassaemia patients in Terengganu are between the ages of 5-19.9 years old (187 of 364 patients, 51.37%).

The majority of these patients are managed by the Paediatric Haemato-Oncologist based at HSNZ. Adult patients in HSNZ are gradually transferred to an adult haematologist.

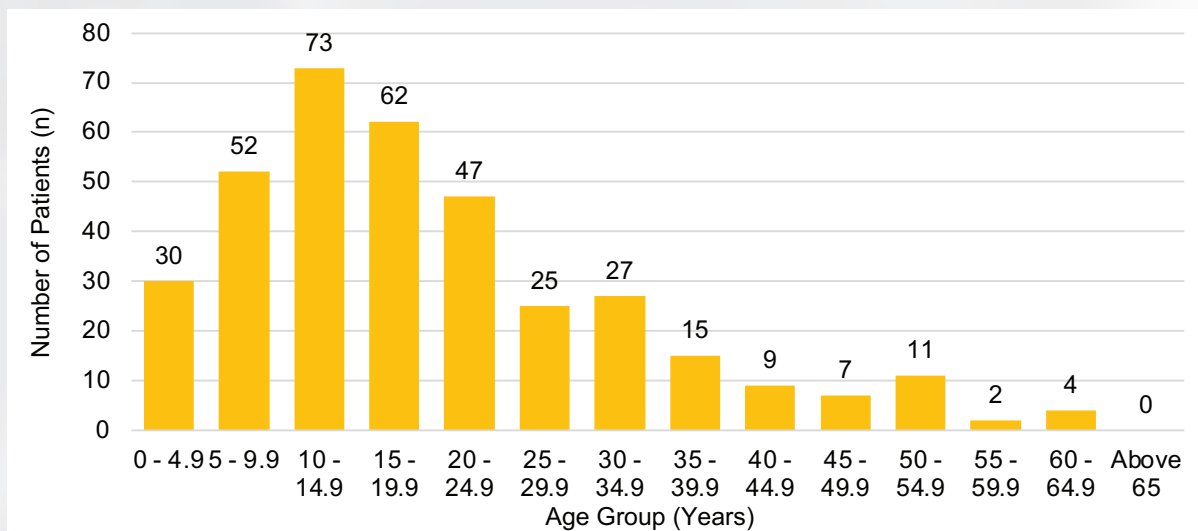


Figure 16.2: Distribution of Patients in Terengganu by Age Group

Table 16.3: Distribution of Patients in Terengganu According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0-14.9	155	β-Thalassaemia Major	24	15.48
		β-Thalassaemia Intermedia	8	5.16
		HbE/β-Thalassaemia	84	54.19
		Hb H Disease	38	24.52
		Others	1	0.65
15-29.9	134	β-Thalassaemia Major	27	20.15
		β-Thalassaemia Intermedia	7	5.22
		HbE/β-Thalassaemia	73	54.48
		Hb H Disease	24	17.91
		Others	3	2.24
30-44.9	51	β-Thalassaemia Major	7	13.73
		β-Thalassaemia Intermedia	8	15.69
		HbE/β-Thalassaemia	32	62.75
		Hb H Disease	3	5.88
		Others	1	1.96
45-59.9	20	β-Thalassaemia Major	2	10.00
		β-Thalassaemia Intermedia	6	30.00
		HbE/β-Thalassaemia	5	25.00
		Hb H Disease	7	35.00
		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
		Hb H Disease	1	25.00
		Others	0	0.00
Total			364	

16.2.2 Gender

There are 174 female patients (47.80%) and 190 male patients (52.20%) in Terengganu. Table 16.4 shows a distribution of the male and female patients by centre.

Table 16.4: Distribution of Patients in Terengganu According to Gender by Centre

Centre	Total	Male		Female	
		No.	%	No.	%
Hospital Sultanah Nur Zahirah	223	117	32.14	106	29.12
Hospital Dungun	24	12	3.30	12	3.30
Hospital Kemaman	35	24	6.59	11	3.02
Hospital Besut	27	14	3.85	13	3.57
Hospital Hulu Terengganu	37	16	4.40	21	5.77
Hospital Setiu	18	7	1.92	11	3.02
Total	364	190	52.20	174	47.80

16.2.3 Ethnic Group

Table 16.5 shows the ethnic distribution of thalassaemia patients in Terengganu. Malay patients form a great majority of patients in the state (356 patients, 97.80%). The ethnic distribution under others include Thais and foreigners.

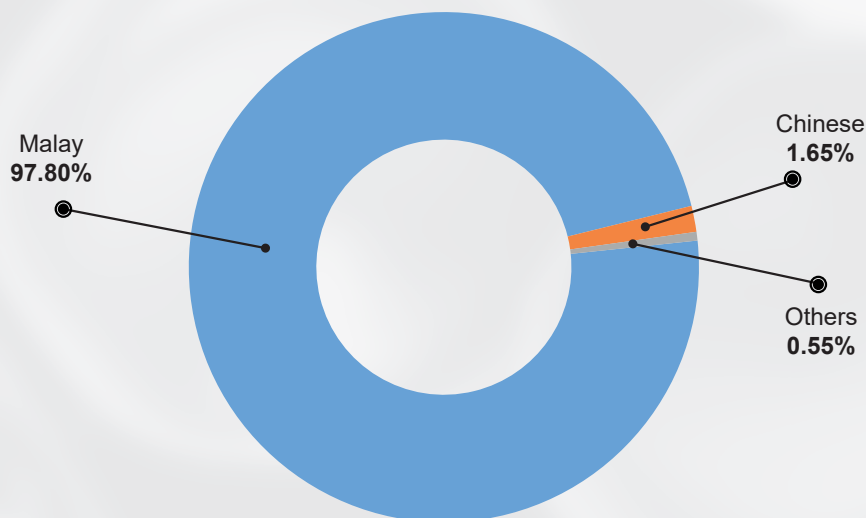


Figure 16.3: Distribution of Patients in Terengganu by Ethnic Group

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

Centre	Total Number of Patients	Malay		Chinese		Indian	
		No.	%	No.	%	No.	%
Hospital Sultanah Nur Zahirah	223	218	59.89	5	1.37	0	0.00
Hospital Hulu Terengganu	37	37	10.16	0	0.00	0	0.00
Hospital Kemaman	35	34	9.34	1	0.27	0	0.00
Hospital Besut	27	26	7.14	0	0.00	1	0.27
Hospital Dungun	24	23	6.32	0	0.00	1	0.27
Hospital Setiu	18	18	4.95	0	0.00	0	0.00
Total	364	356	97.80	6	1.65	2	0.55

16.3 DIAGNOSIS

HbE/ β -thalassaemia formed the major number of the patients (195 patients), followed by Hb H disease (73 patients) and β -thalassaemia major (61 patients).

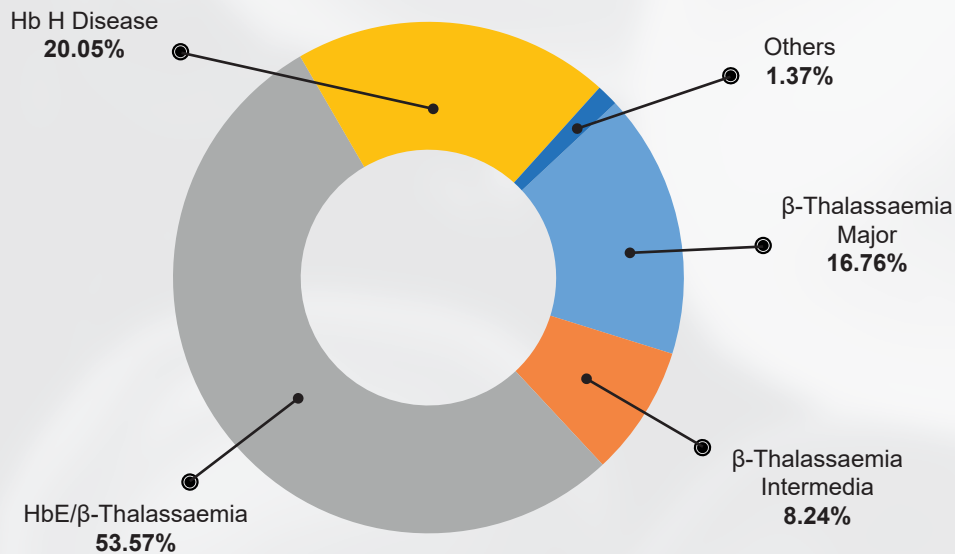


Figure 16.4: Distribution of Patients in Terengganu by Diagnosis

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

Centre	Total Number of Patients	β-Thalassaemia major		β-Thalassaemia intermedia		HbE/β-Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Nur Zahirah	223	24	6.59	19	5.22	128	35.16	49	13.46	3	0.82
Hospital Hulu Terengganu	37	16	4.40	4	1.10	10	2.75	7	1.92	0	0.00
Hospital Kemaman	35	5	1.37	3	0.82	22	6.04	4	1.10	1	0.27
Hospital Besut	27	8	2.20	2	0.55	11	3.02	5	1.37	1	0.27
Hospital Dungun	24	7	1.92	1	0.27	14	3.85	2	0.55	0	0.00
Hospital Setiu	18	1	0.27	1	0.27	10	2.75	6	1.65	0	0.00
Total	364	61	16.76	30	8.24	195	53.57	73	20.05	5	1.37

Table 16.7: 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	61	Malay	59	16.21
		Chinese	0	0.00
		Indian	0	0.00
		Foreigner	1	0.27
		Others	1	0.27
β-Thalassaemia Intermedia	30	Malay	28	7.69
		Chinese	2	0.55
		Indian	0	0.00
		Foreigner	0	0.00
		Others	0	0.00
HbE/β-Thalassaemia	195	Malay	194	53.30
		Chinese	1	0.27
		Indian	0	0.00

		Foreigner	0	0.00
		Others	0	0.00
Hb H Disease	73	Malay	70	19.23
		Chinese	3	0.82
		Indian	0	0.00
		Foreigner	0	0.00
		Others	0	0.00
Others	5	Malay	5	1.37
		Chinese	0	0.00
		Indian	0	0.00
		Foreigner	0	0.00
		Others	0	0.00
Total			364	100.00

16.4 TREATMENT

16.4.1 Iron Chelation

A total of 242 patients in Terengganu were prescribed chelating agents. As shown in Table 16.8, DFP is the most commonly prescribed agent (133 patients), followed by DFX (62 patients) and DFO (23 patients). The most combined therapy prescribed is DFO and DFP (23 patients). A majority of centres in Terengganu commonly prescribed DFP as shown in Table 16.9.

Table 16.8: Distribution of Patients in Terengganu According to Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	23	9.50
DFP only	133	54.96
DFX only	62	25.62
DFO + DFP	23	9.50
DFP + DFX	1	0.41
DFO + DFX	0	0.00
DFO + DFP + DFX	0	0.00
Total	242	100.00

Table 16.9: 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	16	6.61
		DFP only	73	30.17
		DFX only	36	14.88
		DFO + DFP	21	8.68
		DFP + DFX	1	0.41
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Dungun	21	DFO only	1	0.41
		DFP only	15	6.20
		DFX only	4	1.65
		DFO + DFP	1	0.41
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kemaman	24	DFO only	0	0.00
		DFP only	10	4.13
		DFX only	14	5.79
		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	1.24
		DFP only	8	3.31
		DFX only	2	0.83
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Hulu Terengganu	28	DFO only	3	1.24
		DFP only	18	7.44
		DFX only	6	2.48
		DFO + DFP	1	0.41
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Setiu	9	DFO only	0	0.00
		DFP only	9	3.72
		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			242	100.00

Chelating agents are mostly prescribed to patients below 30 years old. DFX is mainly prescribed to patients at ages 0-14.9 years old (23.97%), whereas DFP and DFO was mostly monotherapy prescribed to patients' age 11 years and above.

Table 16.10: 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	98	DFO only	8	8.16
		DFP only	29	29.59
		DFX only	58	59.18
		DFO + DFP	3	3.06
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15-29.9	94	DFO only	11	11.70
		DFP only	68	72.34
		DFX only	1	1.06
		DFO + DFP	14	14.89
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30-44.9	38	DFO only	4	10.53
		DFP only	26	68.42
		DFX only	2	5.26
		DFO + DFP	5	13.16
		DFP + DFX	1	2.63
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-59.9	10	DFO only	0	0.00
		DFP only	9	90.00
		DFX only	0	0.00
		DFO + DFP	1	10.00
		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	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			242	

16.4.2 Serum Ferritin Level

Table 16.11 shows that 189 (75.90%) out of 249 TDT patients have their serum ferritin levels recorded. A total of 118 patients (62.44%) has a serum ferritin level lower than 2,500 ng/mL. The remaining 71 patients (37.56%) have a serum ferritin level higher than 2,500 ng/mL.

Table 16.11: Distribution of TDT 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	138	51	26.98	42	22.22	32	16.93	12	6.35	1	0.53
Hospital Hulu Terengganu	29	4	2.12	10	5.29	9	4.76	6	3.17	0	0.00
Hospital Kemaman	12	2	1.06	5	2.65	4	2.12	1	0.53	0	0.00
Hospital Setiu	4	0	0.00	2	1.06	1	0.53	1	0.53	0	0.00
Hospital Dungun	3	0	0.00	0	0.00	3	1.59	0	0.00	0	0.00
Hospital Besut	3	1	0.53	1	0.53	1	0.53	0	0.00	0	0.00
Total	189	58	30.69	60	31.75	50	26.46	20	10.58	1	0.53

16.5 COMPLICATIONS AND DEATHS

16.5.1 Complications

Table 16.12 shows the distribution of patients according to transfusion transmissible infections. There are 22 cases recorded, namely, 10 cases with anti HCV positive, 10 cases with HCV RNA and two cases with Hepatitis B.

Table 16.12: Distribution of Patient in Terengganu According to Transfusion Transmissible Infections

Infections	2007-2019	2020
Hepatitis C: Anti HCV	10	0
HCV RNA	10	0
Hepatitis B	2	0
HIV	0	0
Total	22	0

Table 16.13 shows that most reported complication related to blood transfusion in thalassaemia patients was hypothyroid with 12 cases. Other than that, seven cases with hypogonadism, four cases with diabetes mellitus, two cases with amenorrhoea, two cases with hypotestosteronemia and one case with hypoestrogenemia.

Table 16.13: Distribution of Patient in Terengganu According to Endocrine Complications

Complications	Number of Patients (n)
Hypothyroid	12
Hypogonadism	7
Diabetes Mellitus	4
Amenorrhoea	2
Hypotestosteronemia	2
Hypoestrogenemia	1
Total	28

16.5.2 Iron Deposition in Heart and Liver Complications

The following Table 16.14 and Table 16.15 show that there were 95 patients who had their liver and cardiac MRI T2* scan results recorded and updated in the registry. About 73 patients (76.8%) have normal grade iron deposition in cardiac and only 6 patients (6.3%) have normal grade iron deposition in liver. Meanwhile, 54 patients (56.8%) have moderate to severe grade of iron deposition in their liver.

Table 16.14: Distribution of Patients in Terengganu According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Nur Zahirah	79	61	64.21	13	13.68	5	5.26	0	0.00
Hospital Hulu Terengganu	11	10	10.53	0	0.00	0	0.00	1	1.05
Hospital Dungun	2	0	0.00	0	0.00	2	2.11	0	0.00
Hospital Setiu	2	2	2.11	0	0.00	0	0.00	0	0.00
Hospital Besut	1	0	0.00	1	1.05	0	0.00	0	0.00
Hospital Kemaman	0	0	0.00	0	0.00	0	0.00	0	0.00
Total	95	73	76.84	14	14.74	7	7.37	1	1.05

Table 16.15: Distribution of Patients in Terengganu According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild /Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Nur Zahirah	79	5	5.26	30	31.58	22	23.16	22	23.16
Hospital Hulu Terengganu	11	1	1.05	2	2.11	5	5.26	3	3.16
Hospital Dungun	2	0	0.00	0	0.00	1	1.05	1	1.05
Hospital Setiu	2	0	0.00	2	2.11	0	0.00	0	0.00
Hospital Besut	1	0	0.00	1	1.05	0	0.00	0	0.00
Hospital Kemaman	0	0	0.00	0	0.00	0	0.00	0	0.00
Total	95	6	6.32	35	36.84	28	29.47	26	27.37

16.5.3 Death Cases

Based on Table 16.16, seven categories of cause of death were reported involving thalassaemia patients in Terengganu. Infections cases were frequently reported with half of the total number. Eight out of 30 cases were related to the cardiac. The least reported causes of death are due to malignancy, liver disease, motor vehicle accident and thrombosis, one case for each category. There were two reported cases of death due to thalassaemia.

Table 16.16: Cumulative Known Cause of Death in Terengganu

Cause of Death	Number of Patients (n)
Infections	16
Cardiac	8
Thalassaemia	2
Malignancy	1
Liver Disease	1
Motor Vehicle Accident (MVA)	1
Thrombosis	1
Total	30

16.6 CONCLUSION

The majority of thalassaemia patients in Terengganu receive treatment at tertiary centre, which is HSNZ (223 out of 364 patients, 61.26%). Patients aged 20 years and below form the majority at 187 patients (51.37%) in Terengganu. Nearly all thalassaemia patients in Terengganu are of Malay ethnicity (356 patients, 97.8%). Male to female patient ratio is 1:1. Based on the current registry data, six new cases were reported in 2020.

There was one death of thalassaemia patient due to infections in 2020, bringing a cumulative total of 30 deaths recorded. Four patients underwent HSCT treatment at IPHKL since 2017. Three of them are considered cured. While one patient is currently under post HSCT treatment. An average of three patients are planned for HSCT per year. HbE/ β -Thalassaemia is the majority diagnosis of the patients (195 patients, 53.57%), followed by Hb H disease (73 patients, 20.05%) and β -Thalassaemia major (61 patients, 16.76%).

DFP is the most prescribed chelating agent, prescribed to 133 patients (54.96%), followed by DFX with 62 patients (25.62%). DFO + DFP is the most common combination of chelating agents prescribed. Patients who are not on chelating agents are mostly NTD patients. In addition, 118 patients (62.44%) of the 249 TDT patients have serum ferritin level lower than 2,500 ng/mL.

Hypothyroid is the highest complication caused by iron overload for patient in Terengganu. Out of 95 patients who had liver and cardiac MRI T2* scan results, about 73 patients have normal grade iron deposition in cardiac but only six patients have normal grade iron deposition in the liver.

17.0

**WILAYAH
PERSEKUTUAN
KUALA LUMPUR**

17.1 INTRODUCTION

The federal territories comprise of three territories, which are Kuala Lumpur, Putrajaya and Labuan. Kuala Lumpur is the capital of Malaysia and has a population of 1.8 million as of 2020 (based on Department of Statistic, Malaysia). The ethnic distribution are Malays (40.49%), Chinese (36.97%), Indian (8.58%), Others (1.14%) and non-Malaysian citizens (12.81%).

There are three major hospitals in Kuala Lumpur that are treating thalassaemia patients, namely, Hospital Tunku Azizah, Pusat Perubatan Universiti Kebangsaan Malaysia and Pusat Perubatan Universiti Malaya.

17.2 PATIENT DEMOGRAPHICS

Patients were categorised as alive, lost to follow-up, cured by transplant or deceased. In 2020, the cumulative total number of patient deaths in Kuala Lumpur are 91 cases. There are 503 patients in Kuala Lumpur, including 97 patients with lost to follow-up and 36 patients with cured by transplant.

Table 17.1: Distribution of Patients in Kuala Lumpur by Centre

Centre	Number of Patients (n)	Percentage (%)
Hospital Tunku Azizah	213	42.35
Pusat Perubatan Universiti Kebangsaan Malaysia	180	35.79
Pusat Perubatan Universiti Malaya	110	21.87
Total	503	100

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

Vital Status	Number of Patients (n)
Alive and On Active Treatment	370
Cured by Stem Cell Therapy	36
Total	406
Lost to Follow Up	97
Total	503
Death in 2020	1
Cumulative Reported Death	91

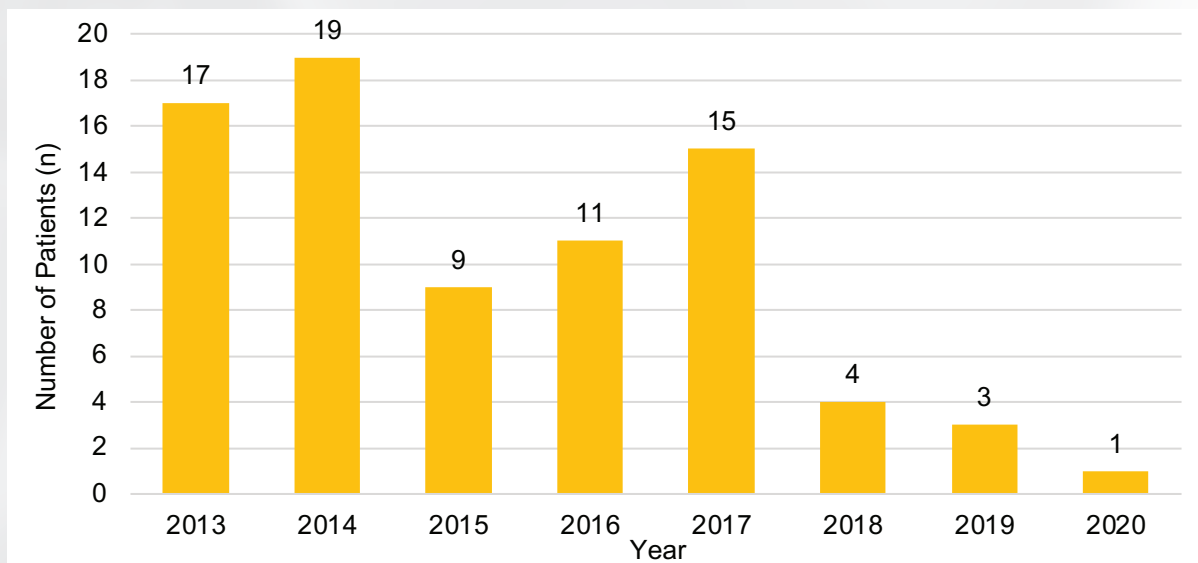


Figure 17.1: Distribution of Thalassaemia Births in Kuala Lumpur by Year

17.2.1 Age Groups

The youngest patient in Kuala Lumpur is 1 year 9 months old and the oldest is 67 years old. Patients aged 5-19.9 years old formed the largest group of patients in Kuala Lumpur. Of these, a sizeable number is transferred to the adult treatment centre once they reach 18 years old. Table 17.3 shows that majority of patients below 45 years old are diagnosed with HbE/ β -thalassaemia.

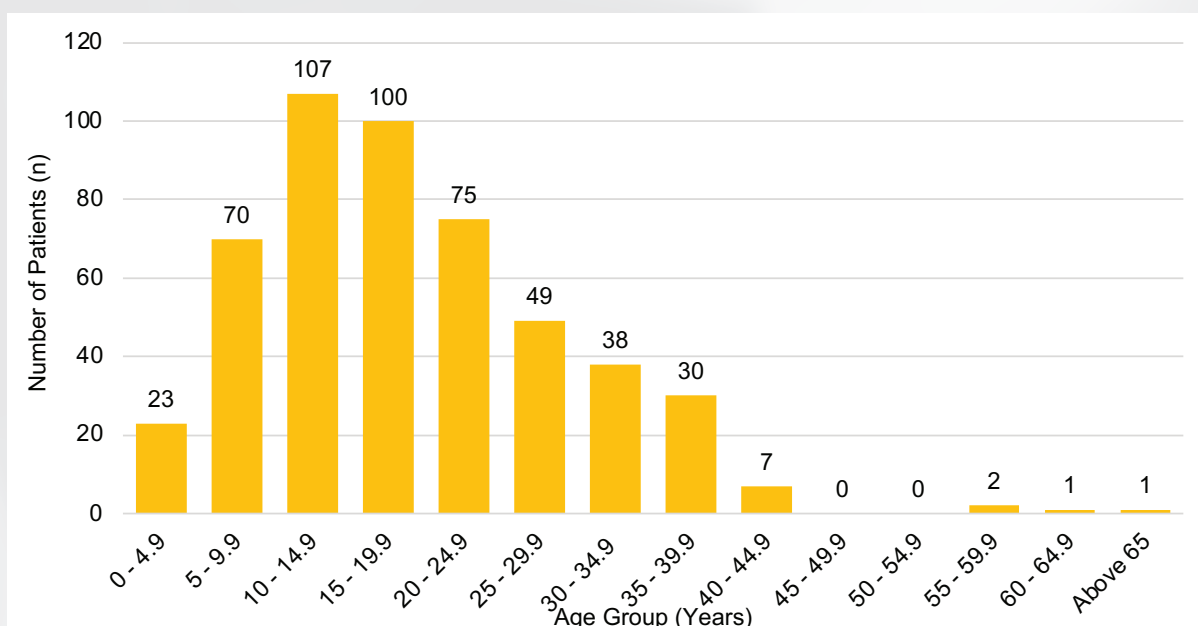


Figure 17.2: Distribution of Patients in Kuala Lumpur by Age Group

Table 17.3: Distribution of Patients in Kuala Lumpur According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0-14.9	200	β-Thalassaemia Major	59	29.50
		β-Thalassaemia Intermedia	3	1.50
		HbE/β-Thalassaemia	82	41.00
		Hb H Disease	42	21.00
		Others	14	7.00
15-29.9	224	β-Thalassaemia Major	53	23.66
		β-Thalassaemia Intermedia	6	2.68
		HbE/β-Thalassaemia	106	47.32
		Hb H Disease	51	22.77
		Others	8	3.57
30-44.9	75	β-Thalassaemia Major	25	33.33
		β-Thalassaemia Intermedia	3	4.00
		HbE/β-Thalassaemia	35	46.67
		Hb H Disease	11	14.67
		Others	1	1.33
45-59.9	2	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	50.00
		HbE/β-Thalassaemia	0	0.00
		Hb H Disease	1	50.00
		Others	0	0.00
60 and above	2	β-Thalassaemia Major	1	50.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	1	50.00
		Hb H Disease	0	0.00
		Others	0	0.00
Total			503	

17.2.2 Gender

The gender distribution of thalassaemia patients in Kuala Lumpur is slightly different. There are 268 male patients (53.28%) and 235 female patients (46.72%) (Table 17.4).

Table 17.4: Distribution of Patients in Kuala Lumpur According to Gender by Centre

Centre	Total	Male		Female	
		No.	%	No.	%
Hospital Tunku Azizah	213	114	22.66	99	19.68
Pusat Perubatan Universiti Kebangsaan Malaysia	180	100	19.88	80	15.9
Pusat Perubatan Universiti Malaya	110	54	10.74	56	11.13
Total	503	268	53.28	235	46.72

17.2.3 Ethnic Group

Malay patients form the majority of 375 patients, followed by the Chinese with 103 patients, Indian with five patients and two patients of Kadazan Dusun. Other groups with 18 patients consisting of Thais (Malaysian), Murut, Bidayuh, foreigner and others are also seeking treatment in Kuala Lumpur. For the non-citizens, the treatment fees and iron chelators need to be paid by foreigners.

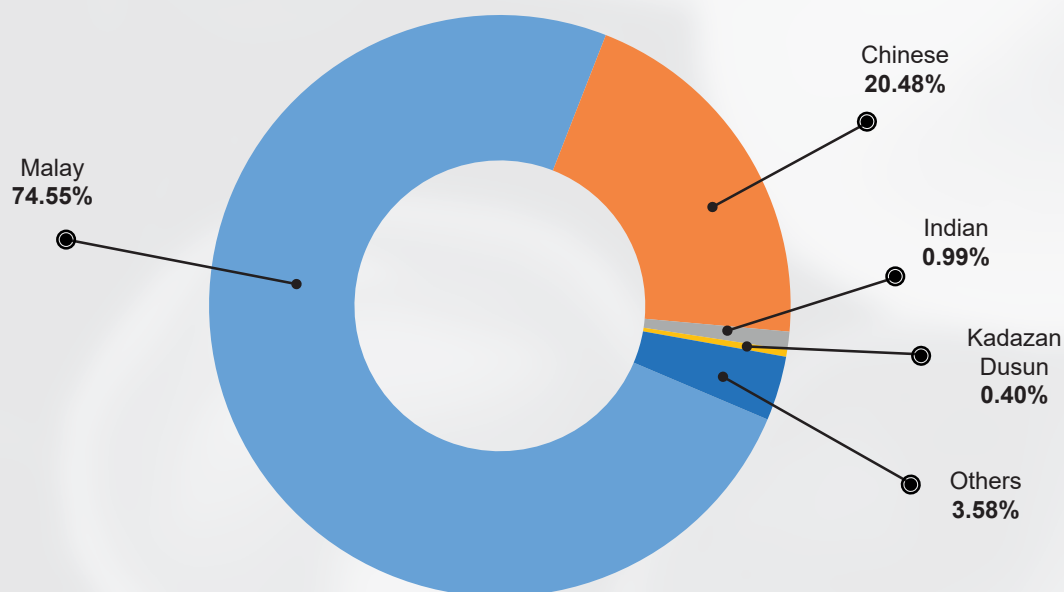


Figure 17.3: Distribution of Patients in Kuala Lumpur by Ethnic Group

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

Centre	Total Number of Patients	Malay		Chinese		Indian		Kadazan Dusun		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Tunku Azizah	213	167	33.20	30	5.96	3	0.60	2	0.40	11	2.19
Pusat Perubatan Universiti Kebangsaan Malaysia	180	137	27.24	39	7.75	0	0.00	0	0.00	4	0.80
Pusat Perubatan Universiti Malaya	110	71	14.12	34	6.76	2	0.40	0	0.00	3	0.60
Total	503	375	74.55	103	20.48	5	0.99	2	0.40	18	3.58

17.3 DIAGNOSIS

Figure 17.4 indicates the diagnoses of HbE/ β -thalassaemia and β -thalassaemia major contributed to 224 patients and 138 patients, respectively. Hb H disease afflicted 105 patients. β -thalassaemia intermedia number contributes to 13 patients and the remaining 23 patients are under other diagnoses.

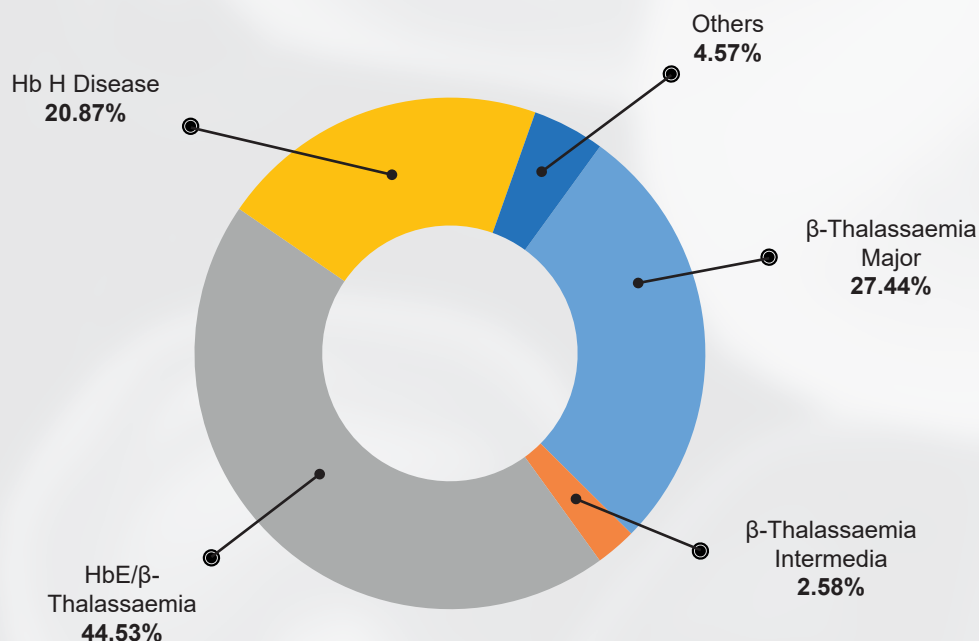


Figure 17.4: Distribution of Patients in Kuala Lumpur by Diagnosis

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

Centre	Total Number of Patients	β -Thalassaemia major		β -Thalassaemia intermedia		HbE/ β -Thalassaemia		Hb H Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Tunku Azizah	213	54	10.74	5	0.99	82	16.30	60	11.93	12	2.39
Pusat Perubatan Universiti Kebangsaan Malaysia	180	33	6.56	6	1.19	95	18.89	38	7.55	8	1.59
Pusat Perubatan Universiti Malaya	110	51	10.14	2	0.40	47	9.34	7	1.39	3	0.60
Total	503	138	27.44	13	2.58	224	44.53	105	20.87	23	4.57

Table 17.7: 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	138	Malay	72	14.31
		Chinese	54	10.74
		Indian	2	0.40
		Kadazan Dusun	1	0.20
		Others	9	1.79
β -Thalassaemia Intermedia	13	Malay	8	1.59
		Chinese	4	0.80
		Indian	1	0.20
		Kadazan Dusun	0	0.00
		Others	0	0.00
HbE/ β -Thalassaemia	224	Malay	198	39.36
		Chinese	15	2.98
		Indian	1	0.20
		Kadazan Dusun	1	0.20
		Others	9	1.79

Hb H Disease	105	Malay	77	15.31
		Chinese	28	5.57
		Indian	0	0.00
		Kadazan Dusun	0	0.00
		Others	0	0.00
Others	23	Malay	20	3.98
		Chinese	2	0.40
		Indian	1	0.20
		Kadazan Dusun	0	0.00
		Others	0	0.00
Total			503	100.00

17.4 TREATMENT

17.4.1 Iron Chelation

Table 17.8 shows that there are 59.44% of TDT and NTD T patients were prescribed iron chelation therapy. About 80.60% of patients are on monotherapy and 19.40% of patients are prescribed a combination of iron chelators. The most prescribed combination chelator is DFO + DFP with 48 patients (16.05%).

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

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	55	18.39
DFP only	40	13.38
DFX only	146	48.83
DFO + DFP	48	16.05
DFP + DFX	3	1.00
DFO + DFX	7	2.34
DFO + DFP + DFX	0	0.00
Total	299	100.00

Table 17.9: 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 (%)
Hospital Tunku Azizah	110	DFO only	8	2.68
		DFP only	1	0.33
		DFX only	94	31.44
		DFO + DFP	1	0.33
		DFP + DFX	0	0.00
		DFO + DFX	6	2.01
		DFO + DFP + DFX	0	0.00
Pusat Perubatan Universiti	93	DFO only	11	3.68
		DFP only	20	6.69
		DFX only	36	12.04
		DFO + DFP	24	8.03
		DFP + DFX	2	0.67
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Pusat Perubatan Universiti Malaya	96	DFO only	36	12.04
		DFP only	19	6.35
		DFX only	16	5.35
		DFO + DFP	23	7.69
		DFP + DFX	1	0.33
		DFO + DFX	1	0.33
		DFO + DFP + DFX	0	0.00
Total			299	100.00

As shown in Table 17.10, the majority of patients below 15 years old received DFX monotherapy. Meanwhile, the 15-29.9 and 30-44.5 years age groups have the highest number of patients who received DFO and DFP combination chelator.

Table 17.10: 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	132	DFO only	14	10.61
		DFP only	5	3.79
		DFX only	105	79.55
		DFO + DFP	2	1.52
		DFP + DFX	0	0.00
		DFO + DFX	6	4.55
		DFO + DFP + DFX	0	0.00
15-29.9	119	DFO only	31	26.05
		DFP only	16	13.45
		DFX only	39	32.77
		DFO + DFP	30	25.21
		DFP + DFX	1	0.84
		DFO + DFX	2	1.68
		DFO + DFP + DFX	0	0.00
30-44.9	44	DFO only	10	22.73
		DFP only	16	36.36
		DFX only	1	2.27
		DFO + DFP	15	34.09
		DFP + DFX	2	4.55
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-59.9	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
60 and above	2	DFO only	0	0.00
		DFP only	1	50.00
		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			299	

17.4.2 Serum Ferritin Level

Table 17.11 shows that there are 179 TDT patients with latest serum ferritin level updated in the registry. There are 41 patients (22.91%) with serum ferritin level lower than 1,000 ng/mL. A total of 60 patients (33.52%) has a serum ferritin level between 1,000-2,500 ng/mL. The number of patients with serum ferritin levels above 2,500 ng/mL are 78 patients (43.57%). The number of patients who are compliant to the iron chelators has slightly increased in 2020.

Table 17.11: Distribution of Patients in Kuala Lumpur According to Serum Ferritin Level in 2020 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 Azizah	105	27	15.08	42	23.46	28	15.64	8	4.47	0	0.00
Pusat Perubatan Universiti Kebangsaan Malaysia	72	14	7.82	16	8.94	26	14.53	11	6.15	5	2.79
Pusat Perubatan Universiti Malaya	2	0	0.00	2	1.12	0	0.00	0	0.00	0	0.00
Total	179	41	22.91	60	33.52	54	30.17	19	10.61	5	2.79

17.5 COMPLICATIONS AND DEATHS

17.5.1 Complications

Based on Table 17.11, there was one patient that are reported with tranfusion transmissible infection of Hepatitis B cummulatively. There was no new tranfusion transmissible infection in 2020. This reflect that patients are receiving safer blood.

Table 17.12: Distribution of Patients in Kuala Lumpur According to Tranfusion Transmissible Infections

Infections	2007-2019	2020
Hepatitis C: Anti HCV	0	0
HCV RNA	0	0
Hepatitis B	1	0
HIV	0	0
Total	1	0

Based on Table 17.13., there are 32 patients with endocrine complications. Short stature contributes the highest number of complications with 22 patients. Meanwhile, five patients are with delayed puberty, two patients with diabetes mellitus, two patients with hypothyroid and one patient with hypocortisol.

Table 17.13: Distribution of Patients in Kuala Lumpur According to Endocrine Complications

Complications	Number of Patients (n)
Short Stature	22
Delayed Puberty	5
Diabetes Mellitus	2
Hypothyroid	2
Hypocortisol	1
Total	32

17.5.2 Iron Deposition in Heart and Liver Complications

There are 52 patients who are performed liver and cardiac MRI T2* scan. Based on table 17.14, there are 33 patients (63.46%) with normal iron loading and five patients (9.62%) with mild iron loading in the cardiac. Moderate and severe iron loading constituted by one patient (1.92%) and 13 patients (25.00%) respectively, most probably due to iron chelation compliance issues.

Based on the Table 17.15, 22 patients (42.31%) with normal iron loading and 17 patients (32.69%) with mild iron loading in their liver. Eleven patients (21.15%) with moderate iron overload and two patients (3.85%) with severe iron loading.

Table 17.14: Distribution of Patients in Kuala Lumpur According to Cardiac MRI T2* by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Pusat Perubatan Universiti Kebangsaan Malaysia	24	21	40.38	1	1.92	1	1.92	1	1.92
Pusat Perubatan Universiti Malaya	16	0	0.00	4	7.69	0	0.00	12	23.08
Hospital Tunku Azizah	12	12	23.08	0	0.00	0	0.00	0	0.00
Total	52	33	63.46	5	9.62	1	1.92	13	25

Table 17.15: Distribution of Patients in Kuala Lumpur According to Liver MRI by Centre

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild /Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Pusat Perubatan Universiti Kebangsaan Malaysia	24	6	11.54	11	21.15	5	9.62	2	3.85
Pusat Perubatan Universiti Malaya	16	16	30.77	0	0.00	0	0.00	0	0.00
Hospital Tunku Azizah	12	0	0.00	6	11.54	6	11.54	0	0.00
Total	52	22	42.31	17	32.69	11	21.15	2	3.85

17.5.3 Death Cases

Based on the Table 17.20, there were 91 patients of cumulative death cases in Kuala Lumpur. Uppermost cause of death is cardiac with 38 patients. Death due to infection recorded with 26 patients. A total of 19 death cases had incomplete data and these patients' causes of death are not included in the table.

Table 17.16: Cumulative Known Cause of Death in Kuala Lumpur

Cause of Death	Number of Patients (n)
Cardiac	38
Infection	26
Died at Home/Brought in Dead to Hospital	2
Motor Vehicle Accident (MVA)	1
Malignancy	1
Central Nervous System Event	1
Endocrine Complications	1
Bone Marrow Transplant Complications	1
Others	1
Total	72

17.6 CONCLUSION

The report describes the outcomes of patient management based on demographic statistics, the types of thalassaemia and effectiveness of treatment. Patients aged between 10–15 years old form the largest age group of patients in Kuala Lumpur. When paediatric patients reach 18 years old, they are transferred to the adult treatment centres which are Hospital Ampang, Pusat Perubatan Universiti Kebangsaan Malaysia or Pusat Perubatan Universiti Malaya. Malay patients are the largest group with 375 patients (74.55%), followed by Chinese with 103 patients (20.48%) and Indians with only five patients (0.99%).

HbE/ β -thalassaemia and β -thalassaemia major afflicted 224 patients (44.53%) and 138 patients (27.44%), respectively. Hb H disease with 105 patients (20.87%), whereas 23 patients (4.57%) with other diagnoses.

In total, 241 patients (80.60%) in Kuala Lumpur were prescribed a monotherapy iron chelator monotherapy and 58 patients (19.40%) were prescribed a combination of iron chelator. A significant number of patients received DFX monotherapy (146 patients, 48.83%). The number of patients who are compliant to iron chelation therapy showed a slight improvement in 2020.

The most common endocrine complications among paediatrics patients are short stature followed by delayed puberty. Out of 52 patients who had liver and cardiac MRI T2* scan done, 22 patients (42.31%) have normal cardiac iron loading and only two patients (3.85%) have severe cardiac iron loading. There are 33 patients (63.46%) with normal liver iron loading and 13 patients (25.00%) with severe liver iron loading. The accessibility to liver and cardiac MRI t2* among thalassaemia patients should be improved.

There were 91 cumulative deaths in Kuala Lumpur included one death reported at Hospital Tunku Azizah due to cardiac cause in 2020. The most common cause of death is due to cardiac with 38 deaths, followed by infection with 26 deaths.

18.0

**WILAYAH
PERSEKUTUAN
LABUAN**

18.1 INTRODUCTION

Labuan is a federal territory in East Malaysia and it is an island located off the coast of Sabah's territory. Based on the Department of Statistics Malaysia, Labuan has a population of 99,300 as at 2019. The population include Bumiputera (76,000, 76.53%), Chinese (9,600, 9.66%), Indian (800, 0.80%), other ethnics (2,900, 2.92%) and non-Malaysian citizens (10,100, 10.17%).

Hospital Labuan has a specialist physician, general surgeon and anesthetic 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 36 thalassaemia patients in Labuan, including one patient who underwent stem cell therapy. The cumulative number of deaths of thalassaemia patients in Labuan are four patients.

Table 18.1: Distribution of Patients in Labuan

Centre	Number of Patients (n)	Percentage (%)
Hospital Labuan	36	100
Total	36	100

Table 18.2: Distribution of Patients in Labuan by Vital Status

Status	Number of Patients (n)
Alive and on Active Treatment	35
Cured by Stem Cell Therapy	1
Total	36
Lost to Follow Up	0
Total	36
Deaths in 2020	0
Cumulative Reported Deaths	4

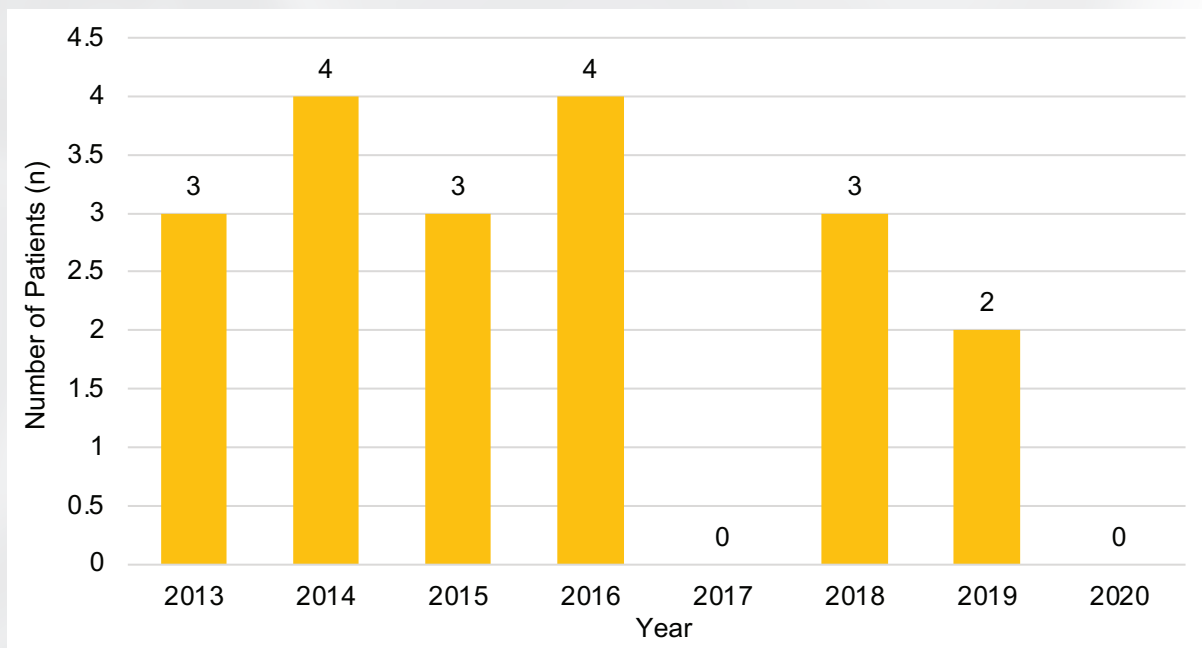


Figure 18.1: Distribution of Thalassaemia Births in Labuan by Year

18.2.1 Age Groups

As shown in Figure 18.2, 34 out of the 36 patients in Labuan are aged below 25 years old. Table 18.3 shows the majority of the patients are diagnosed with β -thalassaemia major.

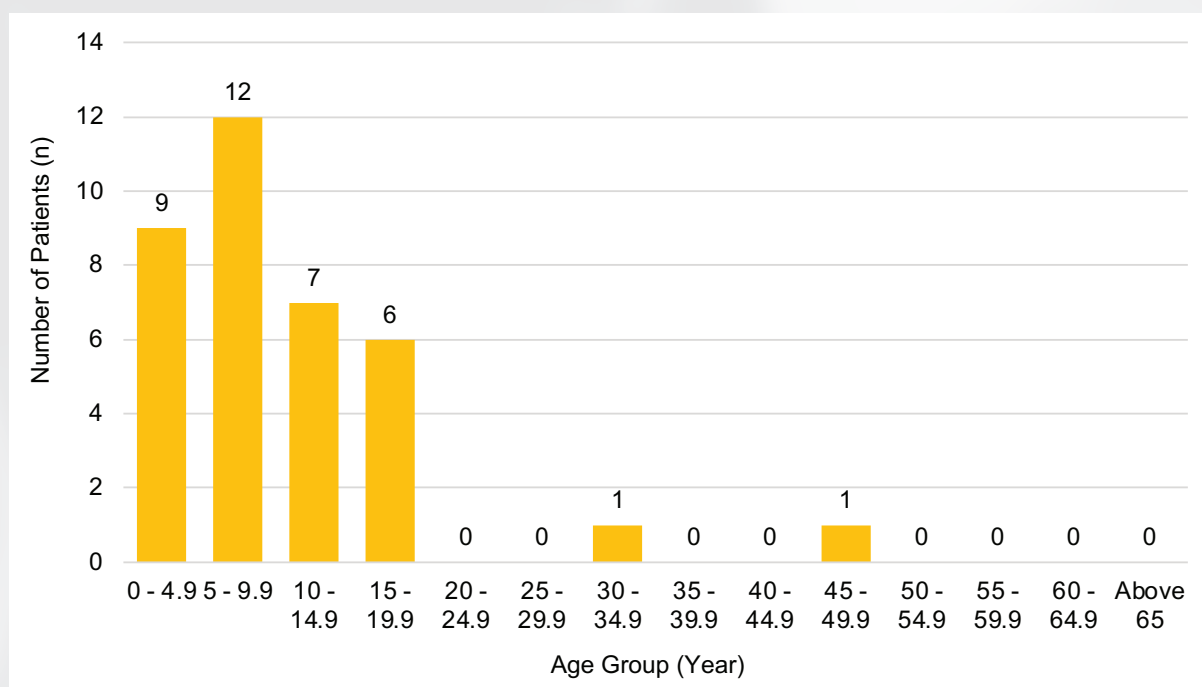


Figure 18.2: Distribution of Patients in Labuan by Age Group

Table 18.3: 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	28	β-Thalassaemia Major	15	41.67
		β-Thalassaemia Intermedia	9	25
		HbE/β-Thalassaemia	0	0.00
		Hb H Disease	4	11.11
		Others	0	0.00
15–29.9	6	β-Thalassaemia Major	5	13.89
		β-Thalassaemia Intermedia	1	2.78
		HbE/β-Thalassaemia	0	0.00
		Hb H Disease	0	0.00
		Others	0	0.00
30–44.9	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	0	0.00
		Hb H Disease	1	2.78
		Others	0	0.00
45 and above	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	2.78
		HbE/β-Thalassaemia	0	0.00
		Hb H Disease	0	0.00
		Others	0	0.00
Total			36	100.00

18.2.2 Gender

Table 18.4 shows the distribution of thalassaemia patients in Labuan by gender. Female patients are higher in number with 21 patients (58.33%) compared to only 15 male patients (41.67%).

Table 18.4: Distribution of Patients in Labuan by Gender

Centre	Total	Male		Female	
		No.	%	No.	%
Hospital Labuan	36	15	41.67	21	58.33
Total	36	15	41.67	21	58.33

18.2.3 Ethnic Group

The Malay form the largest group of patients in Labuan with nine patients (25%), followed by Bajau with eight patients (22%), Kadazan Dusun with seven patients (19%) and Pribumi Sabah six patients (17%). Other ethnicities contribute three patients (8.00%) and finally the Chinese, Murut and Sino Kadazan contribute one patient each (3.00%). The distribution of thalassaemia patients in Labuan according to ethnicity is shown in Figure 18.3.

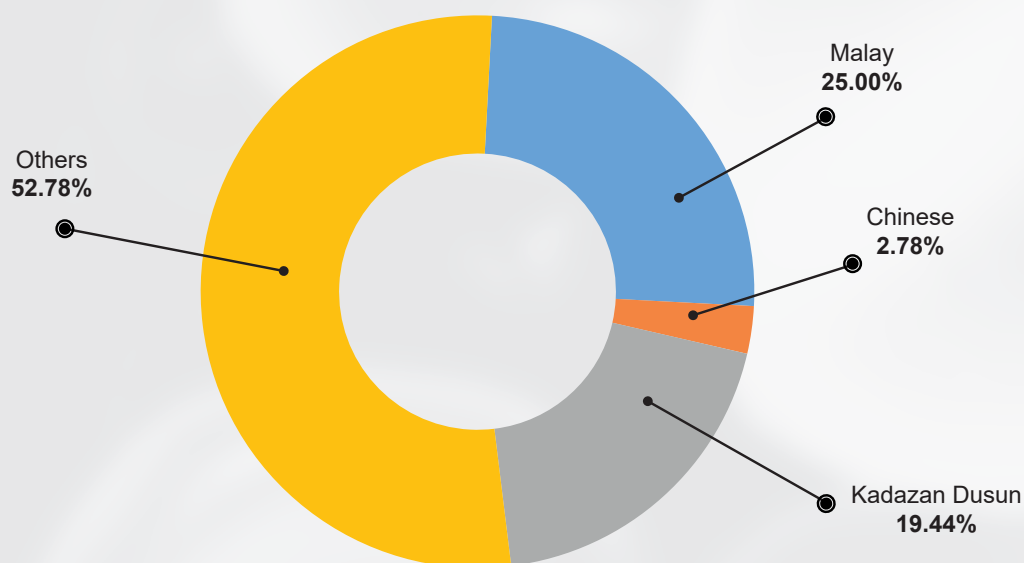


Figure 18.3: Distribution of Patients in Labuan by Ethnic Group

Table 18.5: Distribution of Patients in Labuan According to Ethnic Group

Centre	Total Number of Patients	Malay		Chinese		Indian		Kadazan Dusun		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Labuan	36	9	25.00	1	2.78	7	19.44	19	52.78	11	2.19
Total	36	9	25.00	1	2.78	7	19.44	19	52.78	18	3.58

18.3 DIAGNOSIS

As shown in Figure 18.4, there are 20 patients (55.56%) with β -thalassemia major, followed by 11 patients (30.56%) with β -thalassemia intermedia. The remaining five patients (13.89%) are diagnosed with Hb H disease. Based on Table 18.5, Kadazan Dusun contributes the highest number of β -thalassemia major patients, whereas the Malay has the highest number of β -thalassemia intermedia patients.

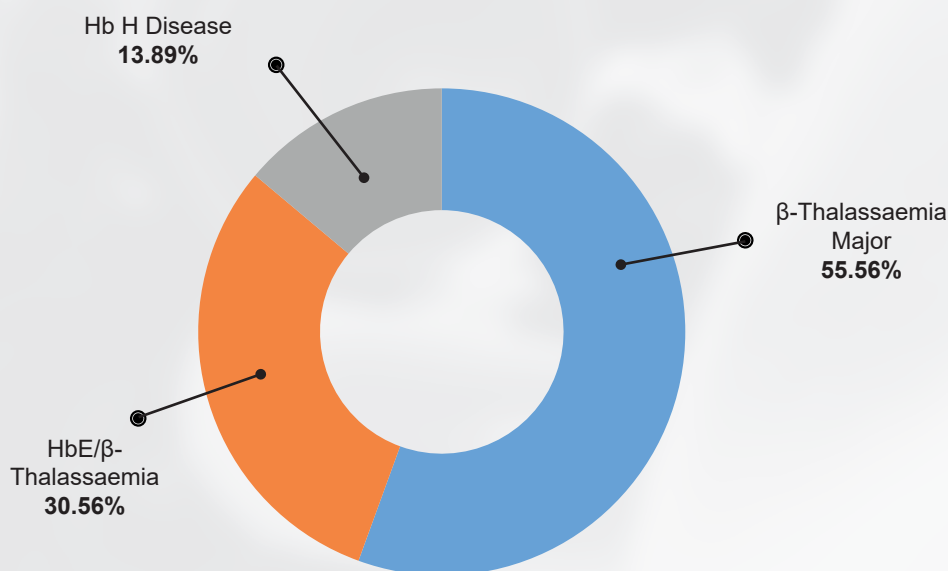


Figure 18.4: Distribution of Patients in Labuan by Diagnosis

Table 18.6: Distribution of Patients in Labuan According to Diagnosis

Centre	Total Number of Patients	β -Thalassaemia major		β -Thalassaemia intermedia		Hb H Disease	
		No.	%	No.	%	No.	%
Hospital Labuan	36	20	55.56	11	30.56	60	11.93
Total	36	20	55.56	11	30.56	105	20.87

Table 18.7: Distribution of Patients in Labuan According to Ethnic Group by Diagnosis

Diagnosis	Total Number of Patients	Ethnicity Group	Number of Patients (n)	Percentage (%)
β-Thalassaemia Major	20	Malay	4	11.11
		Chinese	0	0.00
		Kadazan Dusun	6	16.67
		Others	2	27.78
β-Thalassaemia Intermedia	11	Malay	4	11.11
		Chinese	1	2.78
		Kadazan Dusun	1	2.78
		Others	5	13.89
Hb H Disease	5	Malay	1	2.78
		Chinese	0	0.00
		Kadazan Dusun	0	0.00
		Others	4	11.11
Total			36	100.00

18.4 TREATMENT

18.4.1 Iron Chelation

There are 21 patients who received iron chelation therapy. Ten patients (47.62%) are on DFX. four patients (19.05%) are on DFO followed by 3 patients (14.29%) and 2 patients (9.52%) are prescribed with combination of DFO + DFP and DFP + DFX respectively (Table 18.6). There is 1 patient (4.76%) prescribed DFO + DFP +DFX combination chelator. All patients on DFX monotherapy are below 15 years old and the oldest patient in the age group of 45–59.9 years receive a combination therapy of DFO + DFP.

Table 18.8: Distribution of Patients in Labuan by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	4	19.05
DFP only	1	4.76
DFX only	10	47.62
DFO + DFP	3	14.29
DFP + DFX	2	9.52
DFO + DFX	0	0.00
DFO + DFP + DFX	1	4.76
Total	21	100

Table 18.9: 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	10	76.92
		DFO + DFP	0	0.00
		DFP + DFX	1	7.69
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	7.69
15–29.9	6	DFO only	3	50.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	2	33.33
		DFP + DFX	1	16.67
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30–44.9	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
45 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			21	

18.4.2 Serum Ferritin Level

Based on Table 18.10, 22 patients in Labuan have had their serum ferritin levels measured in 2020. The lowest serum ferritin level recorded is 629.7 ng/mL, and the highest is 5,292 ng/mL. Three patients (13.64%) have serum ferritin level lower than 1,000 ng/mL. Nine patients (40.91%) have serum ferritin levels of between 1,000–2,499 ng/mL, and eight patients (36.36%) had serum ferritin levels of between 2,500–4,999 ng/mL. There are two patients (9.09%) have serum ferritin level between 5,000–9,999 ng/mL.

Table 18.10: 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	22	3	13.64	9	40.91	8	36.36	2	9.09	0	0.00
Total	22	3	13.64	9	40.91	8	36.36	2	9.09	0	0.00

18.5 COMPLICATIONS AND DEATHS

18.5.1 Complications

Up until December 2020, there is no transfusion transmissible infection detected in Labuan indicating that patients are receiving safer blood during treatment.

18.5.2 Iron Deposition in Heart and Liver Complications

There are three patients who had their liver and cardiac MRI T2* scans completed in 2020. Table 18.11 shows that only one patient with moderate iron loading followed by two patients with normal iron loading for cardiac. Based on Table 18.12, there is one patient with severe iron loading and two patients with mild/light iron loading for liver.

Table 18.11: Distribution of Patients in Labuan According to Cardiac MRI T2*

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Labuan	3	2	66.67	0	0.00	1	33.3	0	0.00
Total	3	2	66.67	0	0.00	1	33.3	0	0.00

Table 18.12: Distribution of Patients in Labuan According to Liver MRI

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild /Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Labuan	3	0	0.00	2	66.67	0	0.00	1	33.33
Total	3	0	0.00	2	66.67	0	0.00	1	33.33

18.5.3 Death Cases

Table 18.13 shows that there were 4 deaths reported at Hospital Labuan including 2 patients with unknown cause of death due to unavailable data.

Table 18.13: Cumulative Known Causes of Death in Labuan

Cause of Death	Number of Patients (n)
Infections	1
Bone Marrow Transplant Complications	1
Total	2

18.6 CONCLUSION

This report provides the information regarding the management of patients with thalassemia in Labuan based on the registry. There are nine new thalassaemia patients reported in Labuan in 2020 which makes up a total of 36 thalassaemia patients cumulatively. There were 15 male (41.67%) and 21 female (58.33%) patients in Labuan. The oldest patient is 48 years old whilst the youngest is one year and 7 months old. Malay patients form the largest group with nine patients followed by Bajau with eight patients, Kadazan Dusun with seven patients and several other small ethnics that contribute to the statistics.

In Labuan, β -thalassemia intermedia and β -thalassemia major affected 11 and 20 patients, respectively. The remaining five patients were diagnosed as Hb H disease. From 21 patients that were prescribed chelation therapy, 15 patients (71.42%) in Labuan are on monotherapy and six patients (28.57%) are on combination therapy. The commonest prescribed chelator is DFX with 10 patients (47.62%) and only one patient (4.76%) is prescribed with DFO + DFP + DFX combination chelator. The serum ferritin level was measured by 22 patients in Labuan in 2020. There are only 12 patients (54.55%) who had serum ferritin level lower than 2,500 ng/ml. There was no complication among thalassaemia patients recorded and only four deaths recorded including two cases with unknown cause of death.

19.0

**WILAYAH
PERSEKUTUAN
PUTRAJAYA**

19.1 INTRODUCTION

Putrajaya serves as the federal administrative centre of Malaysia. The population is expected to grow in the relatively new city. Based on Department of Statistic Malaysia, the population is 110,000 in Putrajaya by 2020.

19.2 PATIENT DEMOGRAPHICS

Patients' data were generated based on their vital status, such as active treatment, lost to follow-up or cured by transplant. In Table 19.1, there are 45 alive patients treated in Hospital Putrajaya. 42 patients are on active treatment and another 3 patients have been cured by stem cell therapy.

Table 19.1: Distribution of Patients in Putrajaya

Centre	Number of Patients (n)	Percentage (%)
Hospital Putrajaya	45	100.00
Total	45	100.00

Table 19.2: Distribution of Patients in Putrajaya by Vital Status

Vital Status	Number of Patients (n)
Alive and On Active Treatment	42
Cured by Stem Cell Therapy	3
Total	45
Lost to Follow Up	0
Total	45
Death in 2020	0
Cumulative Reported Death	1

Figure 19.1: Distribution of Thalassaemia Births in Putrajaya by Year

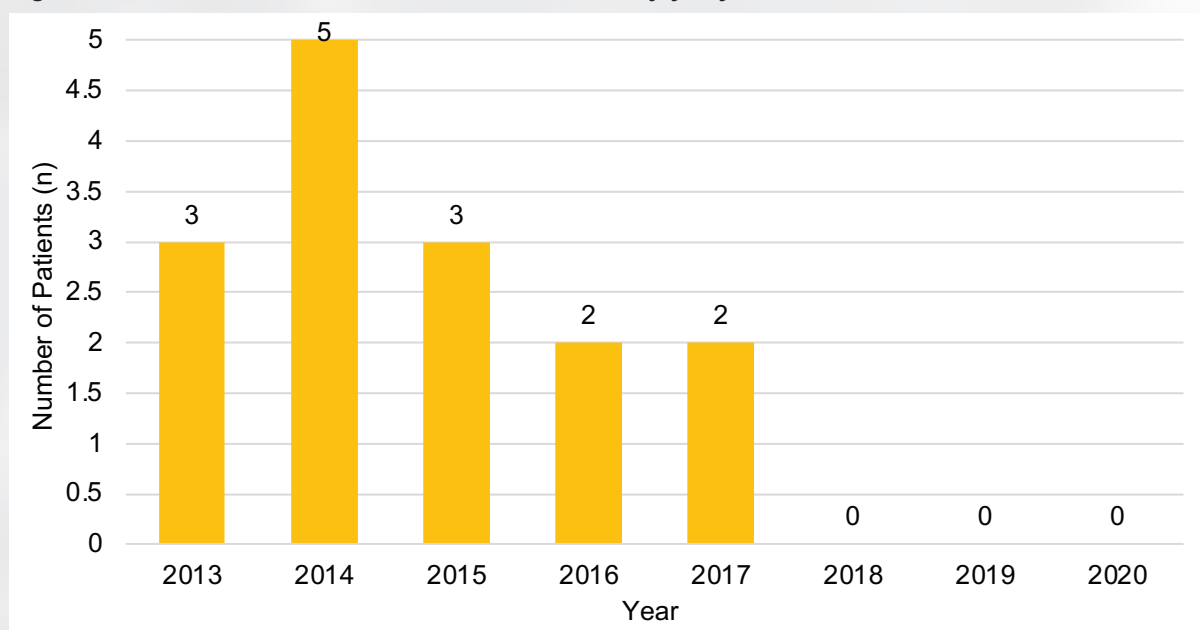


Figure 19.1: Distribution of Thalassaemia Births in Putrajaya by Year

19.2.1 Age Groups

The youngest patient in Putrajaya is three years old and the oldest patient is 46 years old. These patients were diagnosed with β -thalassaemia intermedia. Based on Figure 19.2, the highest number of patients in Putrajaya according to age group is 5–9.9 years old with 18 patients. Based on Table 19.3, most patients were diagnosed with HbE/ β -thalassaemia.

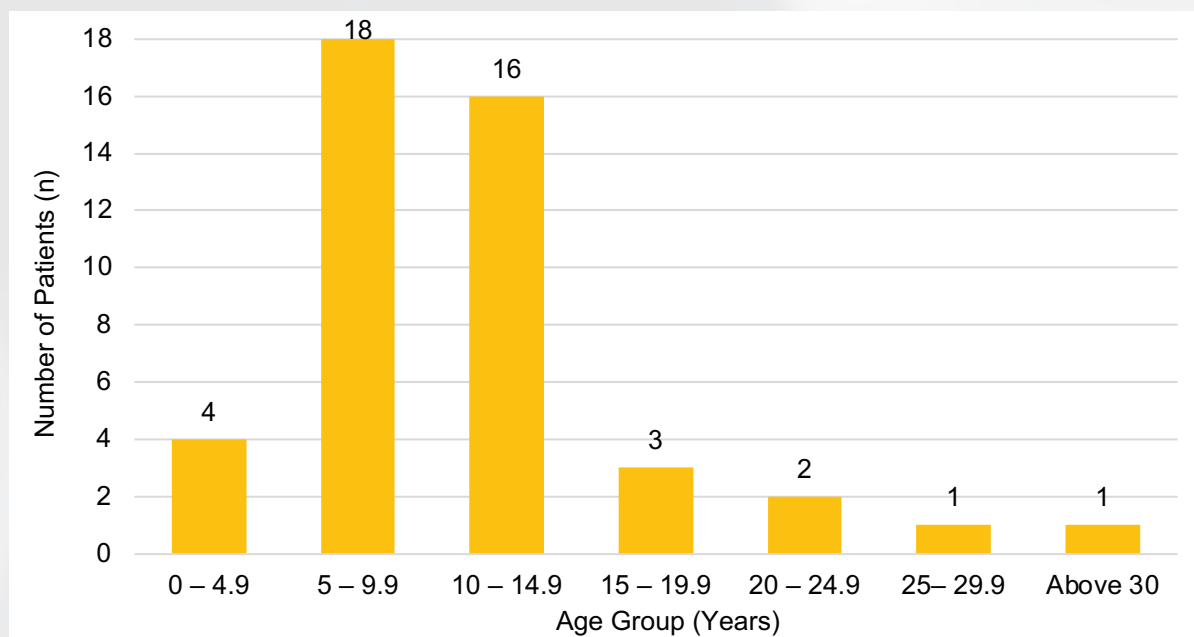


Figure 19.2: Distribution of Patients in Putrajaya by Age Group

Table 19.3: Distribution of Patients in Putrajaya According to Diagnosis by Age Group

Age Group (Years)	Total Number of Patients	Diagnosis	Number of Patient (n)	Percentage (%)
0–14.9	38	β-Thalassaemia Major	8	21.05
		β-Thalassaemia Intermedia	3	7.89
		HbE/β-Thalassaemia	22	57.89
		Hb H Disease	5	13.16
		Others	0	0.00
15–29.9	6	β-Thalassaemia Major	1	16.67
		β-Thalassaemia Intermedia	0	0.00
		HbE/β-Thalassaemia	5	83.33
		Hb H Disease	0	0.00
		Others	0	0.00
30 and above	1	β-Thalassaemia Major	0	0.00
		β-Thalassaemia Intermedia	1	100.00
		HbE/β-Thalassaemia	0	0.00
		Hb H Disease	0	0.00
		Others	0	0.00
Total			45	

19.2.2 Gender

Table 19.4 showing the distribution of patients in Putrajaya by gender. Number of male patients is higher than female patients.

Table 19.4: Distribution of Patients in Putrajaya by Gender

Centre	Total	Male		Female	
		No.	%	No.	%
Hospital Putrajaya	45	33	73.33	12	26.67
Total	45	33	73.33	12	26.67

19.2.3 Ethnic Group

Based on Figure 19.3, thalassaemia patients according to ethnic group in Putrajaya are Malay with 42 patients, two patients are Chinese and one patient is a foreigner.

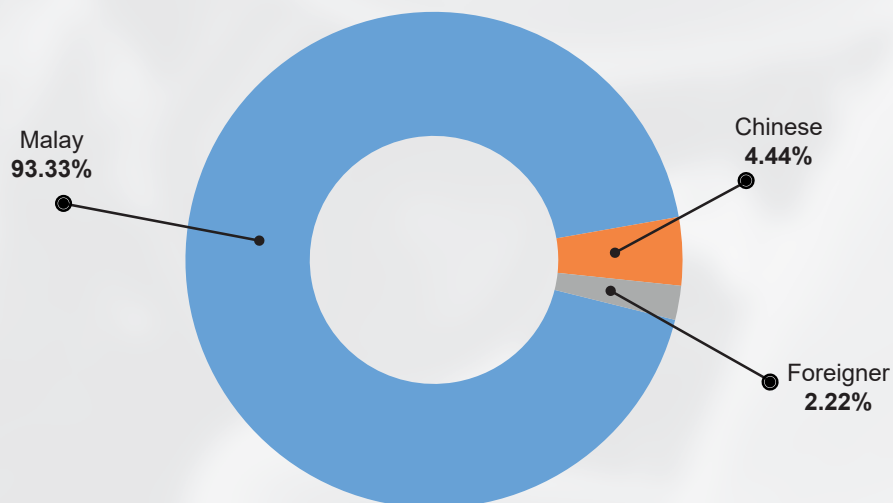


Figure 19.3: Distribution of Patients in Putrajaya by Ethnic Group

Table 19.5: Distribution of Patients in Putrajaya According to Ethnic Group

Centre	Total Number of Patients	Malay		Chinese		Others	
		No.	%	No.	%	No.	%
Hospital Putrajaya	45	42	93.33	2	4.44	1	2.22
Total	45	42	93.33	2	4.44	1	2.22

19.3 DIAGNOSIS

Figure 19.4 shows that more than half of thalassaemia patients in Putrajaya have HbE/ β -thalassaemia with 27 patients (60.00%) and is the majority of diagnosis afflicting Malay ethnic group. Meanwhile, Chinese patients are diagnosed with β -thalassaemia major and Hb H disease.

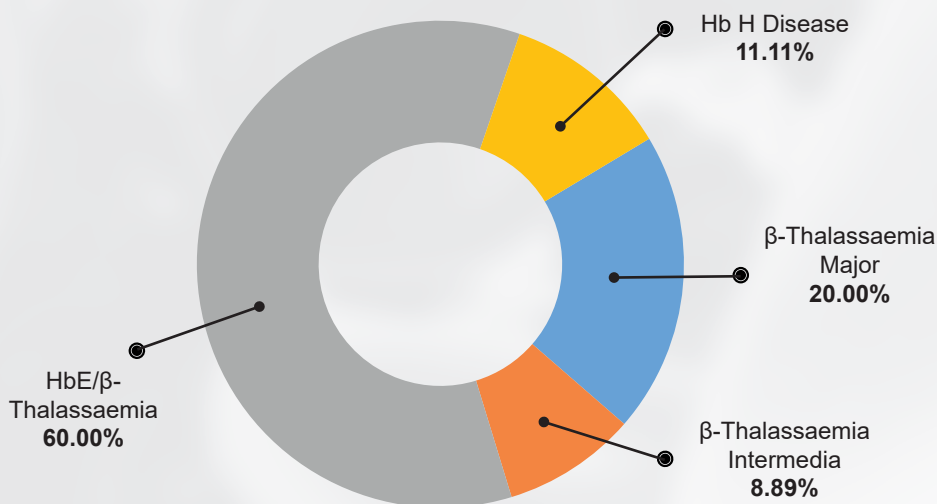


Figure 19.4: Distribution of Patients in Putrajaya by Diagnosis

Table 19.6: Distribution of Patients in Putrajaya According to Diagnosis

Centre	Total Number of Patients	β -Thalassaemia Major		β -Thalassaemia Intermedia		HbE/ β -Thalassaemia		Hb H Disease	
		No.	%	No.	%	No.	%	No.	%
Hospital Putrajaya	45	9	20.00	4	8.89	27	60.00	5	11.11
Total	45	9	20.00	4	8.89	27	60.00	5	11.11

Table 19.7: 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	9	Malay	7	15.56
		Chinese	1	2.22
		Foreigner	1	2.22
β-Thalassaemia Intermedia	4	Malay	4	8.89
HbE/β-Thalassaemia	27	Malay	27	60.00
Hb H Disease	5	Malay	4	8.89
		Chinese	1	2.22
Total			45	100.00

19.4 TREATMENT

19.4.1 Iron Chelationy

Based on Table 19.8, 24 patients are prescribed iron chelator therapy. There are 20 patients (83.33%) who receive DFX, three patients (12.50%) receive DFO and one patient (4.17%) receive DFP monotherapy. Table 19.7 showsng that majority of patient receiving DFX therapy are below 15 years old. The remaining 21 patients in Putrajaya are not prescribed with iron chelation therapy including 16 NTDT patients.

Table 19.8: Distribution of Patients in Putrajaya by Type of Iron Chelator Received

Iron Chelator	Number of Patients (n)	Percentage (%)
DFO only	3	12.50
DFP only	1	4.17
DFX only	20	83.33
DFO + DFP	0	0.00
DFP + DFX	0	0.00
DFO + DFX	0	0.00
DFO + DFP + DFX	0	0.00
Total	24	100.00

Table 19.9: 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	3	15.79
		DFP only	0	0.00
		DFX only	16	84.21
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15–29.9	5	DFO only	0	0.00
		DFP only	1	20.00
		DFX only	4	80.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			24	

19.4.2 Serum Ferritin Level

Based on Table 19.10, 24 patients who received regular transfusion had their serum ferritin levels measured in 2020. Most of these patients have a serum ferritin level below 2,499 ng/mL (19 patients, 79.17%), four patients (16.67%) with serum ferritin level between 2,500–4,999 ng/mL, and one patient (4.17%) with serum ferritin level above 5,000 ng/mL. These results show that 79.17% patients have a serum ferritin level of below 2,499 ng/mL.

Table 19.10: Distribution of TDT 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	24	6	25.00	13	54.17	4	16.67	1	4.17	0	0.00
Total	24	6	25.00	13	54.17	4	16.67	1	4.17	0	0.00

19.5 COMPLICATIONS AND DEATHS

19.5.1 Complications

None of the patients in Putrajaya were reported to suffer any complications of infection or endocrine.

19.5.2 Iron Deposition in Heart and Liver Complications

Based on Table 19.11 and Table 19.12, four patients were tested for their liver and cardiac MRI T2* scan. All patients have normal cardiac iron loading. Meanwhile, there is one TDT patient with HbE/ β -thalassaemia with severe iron loading, one patient with moderate iron loading, one patient with mild iron loading, and one patient with normal iron loading in their liver.

Table 19.11: Distribution of Patients in Putrajaya According to Cardiac MRI T2*

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild/Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Putrajaya	4	4	100.00	0	0.00	0	0.00	0	0.00
Total	4	4	100.00	0	0.00	0	0.00	0	0.00

Table 19.12: Distribution of Patients in Putrajaya According to Liver MRI

Centre	Total Number of Patients	Grade of Iron Deposition							
		Normal		Mild /Light		Moderate		Severe	
		No.	%	No.	%	No.	%	No.	%
Hospital Putrajaya	4	1	25.0	1	25.00	1	25.00	1	25.00
Total	4	1	25.0	1	25.00	1	25.00	1	25.00

19.5.3 Death Cases

Table 19.11 shows that there is only one thalassaemia patient death in Putrajaya and the cause of death is infection.

Table 19.13: Cumulative Known Causes of Death in Putrajaya

Cause of Death	Number of Patients (n)
Infections	1
Total	1

19.6 CONCLUSION

The total number of patients in Putrajaya is 45, and three patients have been cured by stem cell therapy. The number of male patients is higher than female. The majority of the patients are below 15 years of age. The oldest patient is 46 years old in Putrajaya.

The most common type of thalassaemia with the highest number of patients in Putrajaya is HbE/ β -thalassaemia with 27 patients (61.11%) and all are Malay. From 42 patients who are actively receiving treatment in Putrajaya Hospital, 29 patients require regular transfusions. Only 24 TDT patients are prescribed iron chelation therapy and the majority are paediatric patients aged below 18 years old. DFX monotherapy is the most commonly prescribed therapy.

Based on the serum ferritin levels data, 19 TDT patients (79.17%) have serum ferritin levels below 2,500 ng/mL. There were only five patients who had serum ferritin levels above 2,500 ng/mL and most probably they are compliant to the treatment. The liver and cardiac MRI T2* scans were completed for four patients and all of them had normal iron loading for cardiac. The liver MRI scan shows that there is one patient each for severe, moderate, mild and normal liver iron loading. Cardiac and liver iron MRI T2* scan monitoring should be done for more patients for accurate assessment and better management of iron overload among thalassaemia patients.



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