

MALAYSIAN THALASSAEMIA REGISTRY

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FOREWORD

The Malaysian Thalassaemia Registry (MTR) launched on the 12th of May 2007 is part of the National Thalassaemia Prevention and Control programme endorsed by the Malaysian Cabinet in August of 2004. This report is a compilation of work over the past decade showcasing the dynamics of the Malaysian thalassaemia patient landscape after the initiation of a comprehensive national programme. The registry's web-based design allows for real-time data capture and analyses for healthcare personnel in over 100 hospitals nationwide managing thalassaemia patients. Aggregate data made available for policymakers had also enabled better monitoring and planning for the programme at the ministry, state, as well as at the individual hospital level.

Among the most significant finding was the reversal on the survival demography of our transfusion-dependent patients; the majority previously not living beyond the second decade and now with survivorship beyond the fourth decade of life. The enormous healthcare challenge now is on continuing to provide the best of clinical care for an increasingly older cohort of patients while at the same time reducing the number of new births with severe thalassaemia.

The registry has also provided the foundation for research initiatives on many fronts including patient outcomes, laboratory diagnostics, molecular characterisation of local thalassaemia syndromes, quality of life assessments as well as pharmacoeconomic analyses on thalassaemia management in Malaysia. We hope to further improve the wealth of data available in this registry with the largest cohort of thalassaemia patients in the world to serve as a valuable resource of thalassaemia bio-informatics for the country as well as for the wider global thalassaemia fraternity.

The key stakeholders for the MTR are the Medical Development Division and the Ministry's Paediatric Services, both of which provides the much-needed administrative and logistic support. Data collection was accomplished by research assistants supervised by clinicians, with the coordinating centre based in the Paediatric Department, Hospital Kuala Lumpur. The Malaysian Society of Paediatric Haematology & Oncology had also supported the initiative by supplementing grants whenever the need arose.

The initial goal of providing universal iron chelation therapy has been achieved, and in the coming years, provision of optimal and comprehensive care to our increasing number of surviving patients would need to be enhanced further. The preventive aspects of the national programme would need to be revisited and given more emphasis as the number of new births with severe thalassaemia is still significant. It would need a collective effort with adequate resources to address this growing healthcare burden in a sustainable manner.

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Contents

1

Introduction 18

1.1	Thalassaemia	18
1.2	Malaysian Thalassaemia Registry	18
1.3	Demographics of Malaysia	18
1.3.1	Population of Malaysia	19
1.4	Data Collection	19
1.5	Participating Centres	19
1.5.1	Johor	19
1.5.2	Kedah	19
1.5.3	Kelantan	20
1.5.4	Melaka	20
1.5.5	Negeri Sembilan	20
1.5.6	Pahang	20
1.5.7	Perak	20
1.5.8	Perlis	20
1.5.9	Pulau Pinang	20
1.5.10	Sabah	20
1.5.11	Sarawak	21
1.5.12	Selangor	21
1.5.13	Terengganu	21
1.5.14	Wilayah Persekutuan Kuala Lumpur	21
1.5.15	Wilayah Persekutuan Labuan	21
1.5.16	Wilayah Persekutuan Putrajaya	21

2

Registry Report 22

2.1	Ten Centres with the Highest Number of Patients	26
2.2	Patients' Status	26
2.3	Birth Summary	26
2.4	Death Summary	28
2.5	Patient Demographics	32
2.5.1	Age	32
2.5.2	Gender	35
2.5.3	Ethnic group	36
2.6	Diagnosis	40
2.7	Iron Chelation Therapy	44
2.8	Splenectomy	52
2.9	Regular Transfusion	54
2.10	Serum Ferritin Level	56
2.11	Treatment Analysis	59

3

Conclusions and Observations 60

4

Johor 61

4.1	Introduction	61
4.2	Patient Demographics	61
4.2.1	Age	63
4.2.2	Gender	66
4.2.3	Ethnic Group	67

4.3	Diagnosis	69
4.4	Treatment	72
4.4.1	Iron Chelation Therapy	72
4.4.2	Serum Ferritin Level	78
4.5	Observation and Comments	79
4.5.1	Recommendations	79

5 Kedah 80

5.1	Introduction	80
5.2	Patient Demographics	80
5.2.1	Age	82
5.2.2	Gender	85
5.2.3	Ethnic Group	86
5.3	Diagnosis	87
5.4	Treatment	89
5.4.1	Iron Chelation Therapy	89
5.4.2	Serum Ferritin Level	95
5.5	Observation and Comments	96
5.5.1	Recommendations	96
5.5.2	Conclusion	96

6 Kelantan 97

6.1	Introduction	97
6.2	Patient Demographics	97
6.2.1	Age	99
6.2.2	Gender	102
6.2.3	Ethnic Group	103
6.3	Diagnosis	104
6.4	Treatment	106
6.4.1	Iron Chelation Therapy	106
6.4.2	Serum Ferritin Level	112
6.5	Observation and Comments	113
6.5.1	Recommendations	113
6.5.2	Conclusions	113

7 Melaka 114

7.1	Introduction	114
7.2	Patient Demographics	114
7.2.1	Age	115
7.2.2	Gender	118
7.2.3	Ethnic Group	118
7.3	Diagnosis	119
7.4	Treatment	121
7.4.1	Iron Chelation Therapy	121
7.4.2	Serum Ferritin Level	125
7.5	Observation and Comments	125

8 Negeri Sembilan 126

8.1	Introduction	126
8.2	Patient Demographics	126
8.2.1	Age	128
8.2.2	Gender	131
8.2.3	Ethnic Group	131
8.3	Diagnosis	133



9

8.4 Treatment	135
8.4.1 Iron Chelation Therapy	135
8.4.2 Serum Ferritin Level	139
8.5 Observation and Comments	139
8.5.1 Conclusion	140

Pahang 141

9.1 Introduction	141
9.2 Patient Demographics	141
9.2.1 Age	144
9.2.2 Gender	147
9.2.3 Ethnic Group	148
9.3 Diagnosis	149
9.4 Treatment	151
9.4.1 Iron Chelation Therapy	151
9.4.2 Serum Ferritin Level	157
9.5 Observation and Comments	157
9.5.1 Recommendations	158

10

Perak 159

10.1 Introduction	159
10.2 Patient Demographics	159
10.2.1 Age	161
10.2.2 Gender	164
10.2.3 Ethnic Group	165
10.3 Diagnosis	166
10.4 Treatment	169
10.4.1 Iron Chelation Therapy	169
10.4.2 Serum Ferritin Level	174
10.5 Observation and Comments	176
10.5.1 Recommendations	176
10.5.2 Conclusions	176

11

Perlis 177

11.1 Introduction	177
11.2 Patient Demographics	177
11.2.1 Age	178
11.2.2 Gender	181
11.2.3 Ethnic Group	181
11.3 Diagnosis	182
11.4 Treatment	184
11.4.1 Iron Chelation Therapy	184
11.4.2 Serum Ferritin Level	187
11.5 Observation and Comments	188
11.5.1 Recommendations	188
11.5.2 Conclusion	188

12

Pulau Pinang 189

12.1 Introduction	189
--------------------------	------------



12.2	Patient Demographics	189
12.2.1	Age	190
12.2.2	Gender	193
12.2.3	Ethnic Group	194
12.3	Diagnosis	195
12.4	Treatment	197
12.4.1	Iron Chelation Therapy	197
12.4.2	Serum Ferritin Level	202
12.5	Observation and Comments	202
12.5.1	Recommendations	203

13 Sabah 203

13.1	Introduction	203
13.2	Patient Demographics	204
13.2.1	Age	206
13.2.2	Gender	209
13.2.3	Ethnic Group	213
13.3	Diagnosis	214
13.4	Treatment	217
13.4.1	Iron Chelation Therapy	217
13.4.2	Serum Ferritin Level	225
13.5	Observation and Comments	226
13.5.1	Recommendations	226

14 Sarawak 227

14.1	Introduction	227
14.2	Patient Demographics	228
14.2.1	Age	230
14.2.2	Gender	234
14.3	Diagnosis	235
14.4	Treatment	237
14.4.1	Iron Chelation Therapy	237
14.4.2	Serum Ferritin Level	242
14.5	Complications and Deaths	244
14.6	Observation and Comments	246
14.6.1	Recommendations	246

15 Selangor 247

15.1	Introduction	247
15.2	Patient Demographics	247
15.2.1	Age	249
15.2.2	Gender	252
15.2.3	Ethnic Group	253
15.3	Diagnosis	254
15.4	Treatment	257
15.4.1	Iron Chelation Therapy	257
15.4.2	Serum Ferritin Level	263
15.5	Observation and Comments	264
15.5.1	Recommendations	264

16

Terengganu 265

16.1	Introduction	265
16.2	Patient Demographics	265
16.2.1	Age	267
16.2.2	Gender	270
16.2.3	Ethnic Group	270
16.3	Diagnosis	271
16.4	Treatment	274
16.4.1	Iron Chelation Therapy	274
16.4.2	Serum Ferritin Level	278
16.5	Observation and Comments	279
16.5.1	Recommendations	279
16.5.2	Conclusions	279

17

Wilayah Persekutuan Kuala Lumpur 280

17.1	Introduction	280
17.2	Patient Demographics	280
17.2.1	Age	282
17.2.2	Gender	285
17.2.3	Ethnic Group	286
17.3	Diagnosis	287
17.4	Treatment	289
17.4.1	Iron Chelation Therapy	289
17.4.2	Serum Ferritin Level	294
17.4.3	Haemopoietic Transplant in Thalassaemia	294
17.5	Observation and Comments	295

18

Labuan 296

18.1	Introduction	296
18.2	Patient Demographics	296
18.2.1	Age	297
18.2.2	Gender	300
18.2.3	Ethnic Group	300
18.3	Diagnosis	301
18.4	Treatment	303
18.4.1	Iron Chelation Therapy	303
18.4.2	Serum Ferritin Level	307
18.5	Observations and Comments	307
18.5.1	Recommendation	307

19

Putrajaya 308

19.1	Introduction	308
19.2	Patient Demographics	308
19.2.1	Age	309
19.2.2	Gender	311
19.2.3	Ethnic Group	311
19.3	Diagnosis	312
19.4	Treatment	313
19.4.1	Iron Chelation Therapy	313
19.4.2	Serum Ferritin Level	315
19.5	Observation and Comments	316

List of Tables

Table 2.1	: Distribution of Patients in Malaysia by Vital Status	22
Table 2.2	: Number of Thalassaemia Patients by State (2014-2018)	22
Table 2.3	: Top 10 Centres in Malaysia by Number of Thalassaemia Patients	26
Table 2.4	: Number of New Births by State (2011-2018)	27
Table 2.5	: Cumulative Reported Number of Deaths by State until December 2018	29
Table 2.6	: Cumulative Causes of Death Since 2007	29
Table 2.7	: Number of Thalassaemia Patient Deaths by State (2008-2018)	30
Table 2.8	: Distribution of Thalassaemia Patients According to Age Group by State	32
Table 2.9	: Distribution of Patients According to Gender by State	35
Table 2.10	: Distribution of Patients by Ethnic Group	37
Table 2.11	: Distribution of Patients According to Major Ethnic Groups by State	38
Table 2.12	: Distribution of Patients According to Major Ethnic Group Based on Geographical Region	39
Table 2.13	: Distribution of Patients by Diagnosis in 2018	40
Table 2.14	: Distribution of Patients According to Diagnosis by Year	40
Table 2.15	: Distribution of Patients According to Diagnosis by State	41
Table 2.16	: Distribution of Patients According to Diagnosis by Age Group	42
Table 2.17	: Distribution of Patients According to Ethnic Group by Diagnosis	43
Table 2.18	: Distribution of Patients According to Type of Iron Chelator Received	44
Table 2.19	: Distribution of Patients According to Type of Iron Chelator Received by State	46
Table 2.20	: Distribution of Patients According to Type of Iron Chelator Received by Age Group	50
Table 2.21	: Distribution of Splenectomised Patients by State	53
Table 2.22	: Distribution of Patients Receiving Regular Transfusions by State	54
Table 2.23	: Total Number of Patients Receiving Regular Transfusions by Year	56
Table 2.24	: Distribution of Patients According to Most Recent Serum Ferritin Levels by State in 2018	57
Table 2.25	: Distribution of Patients According to Serum Ferritin Levels by Year	57
Table 4.1	: Distribution of Patients by Centre in Johor	61
Table 4.2	: Distribution of Patients by Vital Status in Johor	62
Table 4.3	: Cumulative Causes of Death Since 2007 in Johor	62
Table 4.4	: Distribution of Patients in Johor by Age Group	63
Table 4.5	: Distribution of Patients in Johor According to Diagnosis by Age Group	64
Table 4.6	: Distribution of Patients in Johor by Gender	66
Table 4.7	: Distribution of Patients in Johor by Ethnicity	67
Table 4.8	: Distribution of Patients in Johor According to Ethnic Group by Centre	68
Table 4.9	: Distribution of Patients in Johor by Diagnosis	69
Table 4.10	: Distribution of Patients in Johor According to Diagnosis by Centre	70
Table 4.11	: Distribution of Patients in Johor According to Ethnic Group by Diagnosis	71
Table 4.12	: Distribution of Patients in Johor by Type of Iron Chelator Received	72
Table 4.13	: Distribution of Patients in Johor According to Type of Iron Chelator Received by Centre	73
Table 4.14	: Distribution of Patients in Johor According to Type of Iron Chelator Received by Age Group	75
Table 4.15	: Distribution of Patients in Johor by Most Recent Serum Ferritin Level	78
Table 5.1	: Distribution of Patients in Kedah by Centre	80
Table 5.2	: Distribution of Patients in Kedah by Vital Status	81
Table 5.3	: Cumulative Causes of Death Since 2007 in Kedah	81
Table 5.4	: Distribution of Patients in Kedah by Age Group	82
Table 5.5	: Distribution of Patients in Kedah According to Diagnosis by Age Group	83
Table 5.6	: Distribution of Patients in Kedah by Gender	85
Table 5.7	: Distribution of Patients in Kedah by Ethnic Group	86
Table 5.8	: Distribution of Patients in Kedah According to Ethnic Group by Centre	87
Table 5.9	: Distribution of Patients in Kedah by Diagnosis	87
Table 5.10	: Distribution of Patients in Kedah According to Diagnosis by Centre	88
Table 5.11	: Distribution of Patients in Kedah According to Ethnic Group by Diagnosis	89

List of Tables

Table 5.12	: Distribution of Patients in Kedah by Type of Iron Chelator Received	89
Table 5.13	: Distribution of Patients in Kedah According to Type of Iron Chelator Received by Centre	90
Table 5.14	: Distribution of Patients in Kedah According to Type of Iron Chelator Received by Age Group	92
Table 5.15	: Distribution of Patients in Kedah by Most Recent Serum Ferritin Level	95
Table 6.1	: Distribution of Patients in Kelantan by Centre	97
Table 6.2	: Distribution of Patients in Kelantan by Vital Status	98
Table 6.3	: Cumulative Causes of Death Since 2007 in Kelantan	99
Table 6.4	: Distribution of Patients in Kelantan by Age Group	99
Table 6.5	: Distribution of Patients in Kelantan According to Diagnosis by Age Group	100
Table 6.6	: Distribution of Patients in Kelantan by Gender	102
Table 6.7	: Distribution of Patients in Kelantan by Ethnic Group	103
Table 6.8	: Distribution of Patients in Kelantan According to Ethnic Group by Centre	104
Table 6.9	: Distribution of Patients in Kelantan by Diagnosis	105
Table 6.10	: Distribution of Patients in Kelantan According to Diagnosis by Centre	105
Table 6.11	: Distribution of Patients in Kelantan According to Ethnic Group by Diagnosis	106
Table 6.12	: Distribution of Patients in Kelantan by Type of Iron Chelator Received	107
Table 6.13	: Distribution of Patients in Kelantan According to Type of Iron Chelator Received by Centre	107
Table 6.14	: Distribution of Patients in Kelantan According to Type of Iron Chelator Received by Age Group	110
Table 6.15	: Distribution of Patients in Kelantan by Most Recent Serum Ferritin Level	113
Table 7.1	: Distribution of Patients in Melaka by Centre	114
Table 7.2	: Distribution of Patients in Melaka by Vital Status	115
Table 7.3	: Cumulative Causes of Death Since 2007 in Melaka	115
Table 7.4	: Distribution of Patients in Melaka by Age Group	115
Table 7.5	: Distribution of Patients in Melaka According to Diagnosis by Age Group	116
Table 7.6	: Distribution of Patients in Melaka by Gender	117
Table 7.7	: Distribution of Patients in Melaka by Ethnic Group	118
Table 7.8	: Distribution of Patients in Melaka According to Ethnic Group by Centre	119
Table 7.9	: Distribution of Patients in Melaka by Diagnosis	119
Table 7.10	: Distribution of Patients in Melaka According to Diagnosis by Centre	120
Table 7.11	: Distribution of Patients in Melaka According to Ethnic Group by Diagnosis	120
Table 7.12	: Distribution of Patients in Melaka by Type of Iron Chelator Received	121
Table 7.13	: Distribution of Patients in Melaka According to Type of Iron Chelator Received by Centre	122
Table 7.14	: Distribution of Patients in Melaka According to Type of Iron Chelator Received by Age Group	123
Table 7.15	: Distribution of Patients in Melaka by Most Recent Serum Ferritin Level	125
Table 8.1	: Distribution of Patients in Negeri Sembilan by Centre	126
Table 8.2	: Distribution of Patients in Negeri Sembilan by Vital Status	127
Table 8.3	: Cumulative Causes of Death Since 2007 in Negeri Sembilan	127
Table 8.4	: Distribution of Patients in Negeri Sembilan by Age Group	128
Table 8.5	: Distribution of Patients in Negeri Sembilan According to Diagnosis by Age Group	129
Table 8.6	: Distribution of Patients in Negeri Sembilan According to Gender by Centre	131
Table 8.7	: Distribution of Patients in Negeri Sembilan by Ethnic Group	132
Table 8.8	: Distribution of Patients in Negeri Sembilan According to Ethnic Group by Centre	132
Table 8.9	: Distribution of Patients in Negeri Sembilan by Diagnosis	133
Table 8.10	: Distribution of Patients in Negeri Sembilan According to Diagnosis by Centre	133
Table 8.11	: Distribution of Patients in Negeri Sembilan According to Ethnic Group by Diagnosis	134
Table 8.12	: Distribution of Patients in Negeri Sembilan by Type of Iron Chelator Received	135
Table 8.13	: Distribution of Patients in Negeri Sembilan According to Type of Iron Chelator Received by Centre	135
Table 8.14	: Distribution of Patients in Negeri Sembilan According to Type of Iron Chelator Received by Age Group	137

Table 8.15	: Distribution of Patients in Negeri Sembilan by Most Recent Serum Ferritin Level	139
Table 9.1	: Distribution of Patients in Pahang by Centre	141
Table 9.2	: Distribution of Patients in Pahang by Vital Status	142
Table 9.3	: Cumulative Causes of Death Since 2007 in Pahang	143
Table 9.4	: Number of New Thalassaemia Cases and Births in Pahang by Year	143
Table 9.5	: Distribution of Patients in Pahang by Age Group	144
Table 9.6	: Distribution of Patients in Pahang According to Diagnosis by Age Group	145
Table 9.7	: Distribution of Patients in Pahang by Gender	147
Table 9.8	: Distribution of Patients in Pahang by Ethnic Group	148
Table 9.9	: Distribution of Patients in Pahang According to Ethnic Group by Centre	148
Table 9.10	: Distribution of Patients in Pahang by Diagnosis	149
Table 9.11	: Distribution of Patients in Pahang According to Diagnosis by Centre	150
Table 9.12	: Distribution of Patients in Pahang According to Ethnic Group by Diagnosis	150
Table 9.13	: Distribution of Patients in Pahang by Type of Iron Chelator Received	152
Table 9.14	: Distribution of Patients in Pahang According to Type of Iron Chelator Received by Centre	152
Table 9.15	: Distribution of Patients in Pahang According to Type of Iron Chelator Received by Age Group	154
Table 9.16	: Distribution of Patients in Pahang According to Most Recent Serum Ferritin Level by Centre	157
Table 10.1	: Distribution of Patients in Perak by Centre	159
Table 10.2	: Distribution of Patients in Perak by Vital Status	160
Table 10.3	: Cumulative Causes of Death Since 2007 in Perak	160
Table 10.4	: Distribution of Patients in Perak by Age Group	161
Table 10.5	: Distribution of Patients in Perak According to Diagnosis by Age Group	162
Table 10.6	: Distribution of Patients in Perak by Gender	164
Table 10.7	: Distribution of Patients in Perak by Ethnic Group	165
Table 10.8	: Distribution of Patients in Perak According to Ethnic Group by Centre	166
Table 10.9	: Distribution of Patients in Perak by Diagnosis	166
Table 10.10	: Distribution of Patients in Perak According to Diagnosis by Centre	167
Table 10.11	: Distribution of Patients in Perak According to Ethnic Group by Diagnosis	168
Table 10.12	: Distribution of Patients in Perak by Type of Iron Chelator Received	169
Table 10.13	: Distribution of Patients in Perak According to Type of Iron Chelator Received by Centre	169
Table 10.14	: Distribution of Patients in Perak According to Type of Iron Chelator Received by Age Group	171
Table 10.15	: Distribution of Patients in Perak According to Most Recent Serum Ferritin Level by Centre	174
Table 10.16	: Distribution of TDT Patients in Perak According to Most Recent Serum Ferritin Level by Centre	175
Table 11.1	: Distribution of Patients in Perlis by Centre	177
Table 11.2	: Distribution of Patients in Perlis by Vital Status	177
Table 11.3	: Cumulative Causes of Death Since 2007 in Perlis	177
Table 11.4	: Distribution of Patients in Perlis by Age Group	178
Table 11.5	: Distribution of Patients in Perlis According to Diagnosis by Age Group	179
Table 11.6	: Distribution of Patients in Perlis by Gender	181
Table 11.7	: Distribution of Patients in Perlis by Ethnic Group	181
Table 11.8	: Distribution of Patients in Perlis by Diagnosis	182
Table 11.9	: Distribution of Patients in Perlis According to Ethnic Group by Diagnosis	183
Table 11.10	: Distribution of Patients in Perlis by Type of Iron Chelator Received	184
Table 11.11	: Distribution of Patients in Perlis According to Type of Iron Chelator Received by Age Group	185
Table 11.12	: Distribution of Patients in Perlis According to Most Recent Serum Ferritin Level by Centre	187
Table 12.1	: Distribution of Patients in Pulau Pinang by Centre	189
Table 12.2	: Distribution of Patients in Pulau Pinang by Vital Status	189
Table 12.3	: Cumulative Cause of Death Since 2007 in Pulau Pinang	190
Table 12.4	: Distribution of Patients in Pulau Pinang by Age Group	190
Table 12.5	: Distribution of Patients in Pulau Pinang According to Diagnosis by Age Group	191

List of Tables

Table 12.6	: Distribution of Patients in Pulau Pinang by Gender	193
Table 12.7	: Distribution of Patients in Pulau Pinang by Ethnic Group	194
Table 12.8	: Distribution of Patients in Pulau Pinang According to Ethnic Group by Centre	195
Table 12.9	: Distribution of Patients in Pulau Pinang by Diagnosis	195
Table 12.10	: Distribution of Patients in Pulau Pinang According to Diagnosis by Centre	196
Table 12.11	: Distribution of Patients in Pulau Pinang According to Diagnosis by Ethnic Group	196
Table 12.12	: Distribution of Patients in Pulau Pinang According to Most Recent Serum Ferritin Level by Centre	197
Table 12.13	: Distribution of Patients in Pulau Pinang According to Type of Iron Chelator Received by Centre	198
Table 12.14	: Distribution of Patients in Pulau Pinang According to Type of Iron Chelator Received by Age Group	199
Table 12.15	: Distribution of Patients in Pulau Pinang According to Most Recent Serum Ferritin Level by Centre	202
Table 13.1	: Population of Sabah by Ethnicity	203
Table 13.2	: The Five Administrative Divisions of Sabah and Prevalence of Thalassaemia Patients in These Divisions	203
Table 13.3	: Distribution of Thalassaemia Patients in Each Treatment Centre in Sabah and Estimated Prevalence Rate in the Respective City/Town	204
Table 13.4	: Distribution of Patients in Sabah by Vital Status	205
Table 13.5	: Cumulative Causes of Death Since 2007 in Sabah	205
Table 13.6	: Distribution of Patients in Sabah by Age Group	206
Table 13.7	: Distribution of Patients in Sabah According to Diagnosis by Age Group	207
Table 13.8	: Distribution of Patients in Sabah by Gender	210
Table 13.9	: Distribution of Patients in Sabah by Ethnic Group	211
Table 13.10	: Distribution of Patients in Sabah According to Ethnic Group by Centre	213
Table 13.11	: Distribution of Patients in Sabah by Diagnosis	214
Table 13.12	: Distribution of Patients in Sabah According to Diagnosis by Centre	215
Table 13.13	: Distribution of Patients in Sabah According to Diagnosis by Ethnic Group	215
Table 13.14	: Distribution of Patients in Sabah by Type of Iron Chelator Received	217
Table 13.15	: Distribution of Patients in Sabah According to Type of Iron Chelator Received by Centre	217
Table 13.16	: Distribution of Patients in Sabah According to Type of Iron Chelator Received by Age Group	222
Table 13.17	: Distribution of Patients in Sabah According to Most Recent Serum Ferritin Level by Centre	225
Table 14.1	: Population of Thalassaemia Patients in Sarawak by District	227
Table 14.2	: Distribution of Patients in Sarawak by Centre	228
Table 14.3	: Distribution of Patients in Sarawak by Vital Status	229
Table 14.4	: Distribution of Patients in Sarawak by Age Group	230
Table 14.5	: Distribution of TDT and NTD (Alpha and Beta Thalassaemia) Patients in Sarawak by Age Group	231
Table 14.6	: Distribution of Patients in Sarawak According to Diagnosis by Age Group	232
Table 14.7	: Distribution of Patients in Sarawak According to Gender by Centre	234
Table 14.8	: Distribution of Patients in Sarawak by Diagnosis	235
Table 14.9	: Distribution of Patients in Sarawak According to Diagnosis by Centre	236
Table 14.10	: Distribution of Patients in Sarawak According to Diagnosis by Ethnic Group	236
Table 14.11	: Distribution of Patients in Sarawak by Type of Iron Chelator Received	237
Table 14.12	: Distribution of Patients in Sarawak According to Type of Iron Chelator Received by Centre	238
Table 14.13	: Distribution of Patients in Sarawak According to Type of Iron Chelator Received by Age Group	240
Table 14.14	: Distribution of Patients in Sarawak According to Most Recent Serum Ferritin Level by Centre	242
Table 14.15	: Distribution of TDT Patients in Sarawak According to Most Recent Serum Ferritin Level by Age Group	243

Table 14.16 : Distribution of NTD Patients in Sarawak According to Most Recent Serum Ferritin Level by Age Group	244
Table 14.17 : Distribution of Patients in Sarawak According to Transfusion Status by Centre	244
Table 14.18 : Infectious Complications in Thalassaemia Patients Above 15 Years Old in Sarawak	244
Table 14.19 : Endocrine Complications in Patients Above 15 Years Old in Sarawak Receiving Iron Chelation Therapy	245
Table 14.20 : Cardiac Complication in Patients in Sarawak	245
Table 14.21 : Causes of Death of Patients in Sarawak	245
Table 15.1 : Distribution of Patients in Selangor by Centre	247
Table 15.2 : Distribution of Patients in Selangor by Vital Status	248
Table 15.3 : Cumulative Causes of Death since 1997 in Selangor	248
Table 15.4 : Distribution of Patients in Selangor by Age Group	249
Table 15.5 : Distribution of Patients in Selangor According to Diagnosis by Age Group	250
Table 15.6 : Distribution of Patients in Selangor According to Gender by Centre	252
Table 15.7 : Distribution of Patients in Selangor by Ethnic Group	253
Table 15.8 : Distribution of Patients in Selangor According to Ethnic Group by Centre	254
Table 15.9 : Distribution of Patients in Selangor by Diagnosis	254
Table 15.10 : Distribution of Patients in Selangor According to Diagnosis by Centre	255
Table 15.11 : Distribution of Patients in Selangor According to Ethnic Group by Diagnosis	256
Table 15.12 : Distribution of Patients in Selangor by Type of Iron Chelator Received	257
Table 15.13 : Distribution of Patients in Selangor According to Type of Iron Chelator Received by Centre	257
Table 15.14 : Distribution of Patients in Selangor According to Type of Iron Chelator Received by Age Group	260
Table 15.15 : Distribution of Patients in Selangor According to Most Recent Serum Ferritin Level by Centre in 2018	263
Table 16.1 : Distribution of Patients in Terengganu by Centre	265
Table 16.2 : Distribution of Patients in Terengganu by Vital Status	266
Table 16.3 : Cumulative Causes of Death in Terengganu	266
Table 16.4 : Distribution of Patients in Terengganu by Age Group	267
Table 16.5 : Distribution of Patients in Terengganu According to Diagnosis by Age Group	268
Table 16.6 : Distribution of Patients in Terengganu According to Gender by Centre	270
Table 16.7 : Distribution of Patients in Terengganu by Ethnic Group	270
Table 16.8 : Distribution of Patients in Terengganu According to Ethnic Group by Centre	271
Table 16.9 : Distribution of Patients in Terengganu by Diagnosis	271
Table 16.10 : Distribution of Patients in Terengganu According to Diagnosis by Centre	272
Table 16.11 : Distribution of Patients in Terengganu According to Ethnic Group by Diagnosis	273
Table 16.12 : Distribution of Patients in Terengganu by Type of Iron Chelator Received	274
Table 16.13 : Distribution of Patients in Terengganu According to Type of Iron Chelator Received by Centre	274
Table 16.14 : Distribution of Patients in Terengganu According to Type of Iron Chelator Received by Age Group	276
Table 16.15 : Distribution of Patients in Terengganu According to Most Recent Serum Ferritin Level by Centre	279
Table 17.1 : Distribution of Patients in Kuala Lumpur by Centre	280
Table 17.2 : Distribution of Patients in Kuala Lumpur by Vital Status	281
Table 17.3 : Cumulative Causes of Death Since 1997 in Kuala Lumpur	281
Table 17.4 : Distribution of Patients in Kuala Lumpur by Age Group	282
Table 17.5 : Distribution of Patients in Kuala Lumpur According to Diagnosis by Age Group	283
Table 17.6 : Distribution of Patients in Kuala Lumpur by Gender	285
Table 17.7 : Distribution of Patients in Kuala Lumpur by Ethnic Group	286
Table 17.8 : Distribution of Patients in Kuala Lumpur According to Ethnic Group by Centre	287
Table 17.9 : Distribution of Patients in Kuala Lumpur by Diagnosis	287
Table 17.10 : Distribution of Patients in Kuala Lumpur According to Diagnosis by Centre	288

List of Tables

Table 17.11	: Distribution of Patients in Kuala Lumpur According to Ethnic Group by Diagnosis	288
Table 17.12	: Distribution of Patients in Kuala Lumpur by Type of Iron Chelator Received	289
Table 17.13	: Distribution of Patients in Kuala Lumpur According to Type of Iron Chelator Received by Centre	290
Table 17.14	: Distribution of Patients in Kuala Lumpur According to Type of Iron Chelator Received by Age Group	291
Table 17.15	: Distribution of Patients in Kuala Lumpur According to Serum Ferritin Level in 2017 by Centre	294
Table 17.16	: Distribution of Patients in Kuala Lumpur According to Serum Ferritin Level in 2018 by Centre	294
Table 18.1	: Distribution of Patients in Labuan by Vital Status	296
Table 18.2	: Cumulative Causes of Death Since 2007 in Labuan	296
Table 18.3	: Distribution of Patients in Labuan by Age Group	297
Table 18.4	: Distribution of Patients in Labuan According to Age Group by Diagnosis	298
Table 18.5	: Distribution of Patients in Labuan by Gender	300
Table 18.6	: Distribution of Patients in Labuan by Ethnic Group	300
Table 18.7	: Distribution of Patients in Labuan by Diagnosis	301
Table 18.8	: Distribution of Patients in Labuan According to Ethnic Group by Diagnosis	302
Table 18.9	: Distribution of Patients in Labuan by Type of Iron Chelator Received	303
Table 18.10	: Distribution of Patients in Labuan According to Type of Iron Chelator Received by Age Group	304
Table 18.11	: Distribution of Patients in Labuan According to Most Recent Serum Ferritin Level	307
Table 19.1	: Distribution of Patients in Putrajaya by Centre	308
Table 19.2	: Distribution of Patients in Putrajaya by Vital Status	308
Table 19.3	: Cumulative Cause of Death Since 1997 in Putrajaya	308
Table 19.4	: Distribution of Patients in Putrajaya by Age Group	309
Table 19.5	: Distribution of Patients in Putrajaya According to Diagnosis by Age Group	310
Table 19.6	: Distribution of Patients in Putrajaya by Gender	311
Table 19.7	: Distribution of Patients in Putrajaya by Ethnic Group	311
Table 19.8	: Distribution of Patients in Putrajaya by Ethnic Group by Centre	312
Table 19.9	: Distribution of Patients in Putrajaya by Diagnosis	312
Table 19.10	: Distribution of Patients in Putrajaya According to Ethnic Group by Diagnosis	313
Table 19.11	: Distribution of Patients in Putrajaya by Type of Iron Chelator Received	313
Table 19.12	: Distribution of Patients in Putrajaya According to Type of Iron Chelator Received by Age Group	314
Table 19.13	: Distribution of Patients in Putrajaya According to Most Recent Serum Ferritin Level in 2018	315

List of Figures

Figure 2.1	: Total Number of Thalassaemia Patients by State in 2018	23
Figure 2.2	: Total Number of Patients by Year (2007-2018)	24
Figure 2.3	: Total Number of Patients by State (2007-2018)	25
Figure 2.4	: Total Number of New Births (2011-2018)	27
Figure 2.5	: Number of New Births by State (2013-2018)	28
Figure 2.6	: Total Number of Deaths by Year (2008-2018)	30
Figure 2.7	: Total Number of Deaths by State (2008-2018)	31
Figure 2.8	: Total Number of Patients by Age Group	33
Figure 2.9	: Distribution of Patients According to Age Group by State	34
Figure 2.10	: Distribution of Patients by Gender	35
Figure 2.11	: Distribution of Patients According to Gender by State	36
Figure 2.12	: Distribution of Patients by Ethnic Group	37
Figure 2.13	: Distribution of Patients According to Major Ethnic Groups by Geographical Region	39
Figure 2.14	: Distribution of Patients by Diagnosis	41
Figure 2.15	: Distribution of Patients According to Diagnosis by State	42
Figure 2.16	: Percentage Breakdown of Diagnoses by Ethnic Group	43
Figure 2.17	: Distribution of Patients According to Type of Iron Chelator Received	45
Figure 2.18	: Distribution of Patients According to Type of Iron Chelator Received by State in 2017	45
Figure 2.19	: Distribution of Patients According to Type of Iron Chelator Received by State	49
Figure 2.20	: Distribution of Splenectomised Patients by State	53
Figure 2.21	: Distribution of Patients Receiving Regular Transfusions by State	55
Figure 2.22	: Total Number of Transfusions by State	55
Figure 2.23	: Distribution of Patients According to Serum Ferritin Levels by Year	58
Figure 2.24	: Treatment Analysis	59
Figure 4.1	: Distribution of Patients by Centre in Johor	62
Figure 4.2	: Distribution of New Thalassaemia Births in Johor, According to Type of Thalassaemia by Year	63
Figure 4.3	: Distribution of Patients in Johor by Age Group	64
Figure 4.4	: Distribution of Patients in Johor by Gender	67
Figure 4.5	: Distribution of Patients in Johor by Ethnic Group	67
Figure 4.6	: Distribution of Patients in Johor According to Ethnic Group by Centre	68
Figure 4.7	: Distribution of Patients in Johor by Diagnosis	69
Figure 4.8	: Distribution of Patients in Johor According to Type of Thalassaemia by Age Group	70
Figure 4.9	: Distribution of Patients in Johor by Type of Iron Chelator Received	72
Figure 4.10	: Distribution of Patients in Johor According to Type of Iron Chelator Received by Age Group	78
Figure 5.1	: Distribution of Patients in Kedah by Centre	80
Figure 5.2	: Distribution of Patients in Kedah by Age Group	82
Figure 5.3	: Distribution of Patients in Kedah by Gender	85
Figure 5.4	: Distribution of Patients in Kedah by Ethnic Group	86
Figure 5.5	: Distribution of Patients in Kedah by Diagnosis	88
Figure 5.6	: Distribution of Patients in Kedah by Type of Iron Chelator Received	92
Figure 6.1	: Distribution of Patients in Kelantan by Centre	98
Figure 6.2	: Distribution of Patients in Kelantan by Age Group	100
Figure 6.3	: Distribution of Patients in Kelantan by Gender	103
Figure 6.4	: Distribution of Patients in Kelantan by Ethnic Group	104
Figure 6.5	: Distribution of Patients in Kelantan by Diagnosis	105
Figure 6.6	: Distribution of Patients in Kelantan by Type of Iron Chelator Received	109
Figure 7.1	: Distribution of Patients in Melaka by Centre	114
Figure 7.2	: Distribution of Patients in Melaka by Age Group	116
Figure 7.3	: Distribution of Patients in Melaka by Gender	118
Figure 7.4	: Distribution of Patients in Melaka by Ethnic Group	119

List of Figures

Figure 7.5	: Distribution of Patients in Melaka by Diagnosis	120
Figure 7.6	: Distribution of Patients in Melaka by Type of Iron Chelator Received	122
Figure 8.1	: Distribution of Patients in Negeri Sembilan by Centre	127
Figure 8.2	: Distribution of Patients in Negeri Sembilan by Age Group	128
Figure 8.3	: Distribution of Patients in Negeri Sembilan by Gender	131
Figure 8.4	: Distribution of Patients in Negeri Sembilan by Ethnic Group	132
Figure 8.5	: Distribution of Patients in Negeri Sembilan by Diagnosis	133
Figure 8.6	: Distribution of Patients in Negeri Sembilan by Type of Iron Chelator Received	136
Figure 9.1	: Distribution of Patients in Pahang by Centre	142
Figure 9.2	: Distribution of Patients in Pahang by Age Group	144
Figure 9.3	: Distribution of Patients in Pahang by Gender	147
Figure 9.4	: Distribution of Patients in Pahang by Ethnic Group	148
Figure 9.5	: Distribution of Patients in Pahang by Diagnosis	149
Figure 9.6	: Distribution of Alpha Thalassaemia Patients in Pahang by Ethnicity	151
Figure 9.7	: Distribution of Beta Thalassaemia Patients in Pahang by Ethnicity	151
Figure 9.8	: Distribution of Patients by Type of Iron Chelator Received	154
Figure 10.1	: Distribution of Patients in Perak by Centre	160
Figure 10.2	: Distribution of Patients in Perak by Age Group	161
Figure 10.3	: Distribution of Patients in Perak by Gender	164
Figure 10.4	: Distribution of Patients in Perak by Ethnic Group	165
Figure 10.5	: Distribution of Patients in Perak by Diagnosis	167
Figure 10.6	: Distribution of Patients in Perak by Type of Iron Chelator Received	171
Figure 10.7	: Distribution of Patients in Perak by Most Recent Serum Ferritin Level	175
Figure 11.1	: Distribution of Patients in Perlis by Age Group	178
Figure 11.2	: Distribution of Patients in Perlis by Gender	181
Figure 11.3	: Distribution of Patients in Perlis by Ethnic Group	182
Figure 11.4	: Distribution of Patients in Perlis by Diagnosis	183
Figure 11.5	: Distribution of Patients in Perlis by Type of Iron Chelator Received	184
Figure 12.1	: Distribution of Patients in Pulau Pinang by Centre	189
Figure 12.2	: Distribution of Age Patients in Pulau Pinang by Age Group	191
Figure 12.3	: Distribution of Patients in Pulau Pinang by Gender	193
Figure 12.4	: Distribution of Patients in Pulau Pinang by Ethnic Group	194
Figure 12.5	: Distribution of Population of Pulau Pinang by Ethnic Group	195
Figure 12.6	: Distribution of Patients in Pulau Pinang by Diagnosis	196
Figure 12.7	: Distribution of Patients in Pulau Pinang by Type of Iron Chelator Received	198
Figure 13.1	: Distribution of Patients in Sabah by Centre	205
Figure 13.2	: Distribution of Patients in Sabah by Age Group	207
Figure 13.3	: Distribution of Patients in Sabah by Gender	209
Figure 13.4	: Distribution of Patients in Sabah by Ethnic Group	212
Figure 13.5	: Distribution of Patients in Sabah by Ethnic Sub-Group	212
Figure 13.6	: Distribution of Patients in Sabah by Diagnosis	214
Figure 13.7	: Distribution of Patients in Sabah by Type of Iron Chelator Received	222
Figure 14.1	: Distribution of Patients in Sarawak by Centre	229
Figure 14.2	: New Thalassaemia Births in Sarawak by Year	229
Figure 14.3	: Distribution of Patients in Sarawak by Age Group	230
Figure 14.4	: Distribution of TDT and NTDT (Alpha and Beta Thalassaemia) Patients in Sarawak by Age Group	231
Figure 14.5	: Distribution of Patients in Sarawak by Gender	234
Figure 14.6	: Distribution of Patients in Sarawak by Diagnosis	235
Figure 14.7	: Distribution of Patients in Sarawak by Type of Iron Chelator Received	239
Figure 14.8	: Distribution of Patients in Sarawak According to Most Recent Serum Ferritin Level	243



Figure 15.1 : Distribution of Patients in Selangor by Centre	248
Figure 15.2 : Distribution of Patients in Selangor by Age Group	249
Figure 15.3 : Distribution of Patients in Selangor by Gender	252
Figure 15.4 : Distribution of Patients in Selangor by Ethnic Group	253
Figure 15.5 : Distribution of Patients in Selangor by Diagnosis	255
Figure 15.6 : Distribution of Patients in Selangor by Type of Iron Chelator Received	260
Figure 16.1 : Distribution of Patients in Terengganu by Centre	266
Figure 16.2 : Distribution of Patients in Terengganu by Age Group	267
Figure 16.3 : Distribution of Patients in Terengganu by Gender	270
Figure 16.4 : Distribution of Patients in Terengganu by Ethnic Group	271
Figure 16.5 : Distribution of Patients in Terengganu by Diagnosis	272
Figure 16.6 : Distribution of Patients in Terengganu by Type of Iron Chelator Received	275
Figure 17.1 : Distribution of Patients in Kuala Lumpur by Centre	281
Figure 17.2 : Distribution of Patients in Kuala Lumpur by Age Group	282
Figure 17.3 : Distribution of Patients in Kuala Lumpur by Gender	285
Figure 17.4 : Distribution of Patients in Kuala Lumpur by Ethnic Group	286
Figure 17.5 : Distribution of Patients in Kuala Lumpur by Diagnosis	288
Figure 17.6 : Distribution of Patients in Kuala Lumpur by Type of Iron Chelator Received	290
Figure 17.7 : Number of Thalassaemia Patients Who Underwent Stem Cell Transplant at IPHKL by Year	294
Figure 18.1 : Distribution of Patients in Labuan by Age Group	297
Figure 18.2 : Distribution of Patients in Labuan by Gender	300
Figure 18.3 : Distribution of Patients in Labuan by Ethnic Group	301
Figure 18.4 : Distribution of Patients in Labuan by Diagnosis	302
Figure 18.5 : Distribution of Patients in Labuan by Type of Iron Chelator Received	304
Figure 19.1 : Distribution of Patients in Putrajaya by Age Group	309
Figure 19.2 : Distribution of Patients in Putrajaya by Gender	311
Figure 19.3 : Distribution of Patients in Putrajaya by Ethnic Group	312
Figure 19.4 : Distribution of Patients in Putrajaya by Diagnosis	313
Figure 19.5 : Distribution of Patients in Putrajaya by Type of Iron Chelator Received	314

1 Introduction

1.1 Thalassaemia

Thalassaemia is the most common autosomal recessive disorder involving abnormal haemoglobin formation, with high prevalence in tropical countries including Malaysia. The reduced production or abnormality in one or more of the four globin subunits of haemoglobin would result in thalassaemia. It is estimated that about five percent of Malaysians are carriers, with the majority having alpha or beta thalassaemia with compound heterozygosity with other haemoglobinopathies.

Beta thalassaemia major produces severe anaemia that requires life-long blood transfusion for survival. The molecular defects producing beta thalassaemia are heterogeneous with each ethnic group having unique mutations. Alpha-thalassaemia major is also not uncommon. The fatal form of the condition Hb Bart's hydrops fetalis, is present mainly among the Malaysian Chinese. Thalassaemia is more prominent among the Malay and Kadazan-Dusun ethnicities and it has rarely been observed in the Malaysian Indians. There is very little information regarding thalassaemia in the indigenous population of Peninsular Malaysia.

Regular blood transfusions are the mainstay of treatment for the thalassaemia major patients. The treatment results in transfusional iron overload which requires expensive iron chelation therapy. Unbridled chronic tissue iron deposition would result in hepatic dysfunction and failure, endocrinopathies, cardiac dysfunction and premature death. Clinical datasets documenting the trajectory of thalassaemia patients in the country would assist in its management as well as provide valuable insight on patient outcome and the national healthcare burden for this life-long chronic disorder.

1.2 Malaysian Thalassaemia Registry

The Malaysian Thalassaemia Registry (MTR) is a national registry for all thalassaemia patients who received treatment at hospitals under the Ministry of Health Malaysia and three main university hospitals. This registry provides comprehensive updated information on thalassaemia patients, including their socio-demographic, clinical records, type of treatment received, death records and other complications collated over the years, to give an overview of thalassaemia prevalence and incidence across the country as well as its clinical outcome.

The registry is a web-based system accessed by different users who have been enrolled through the website portal: www.mytalasemia.net.my

MyTalasemia is a portal that takes a holistic approach in its implementation recognising that managing thalassaemia requires concerted action by all stakeholders including patients, medical professionals, the public and the government.

1.3 Demographics of Malaysia

Malaysia consists of thirteen states and three federal territories. Eleven states and two federal territories are located on the Malay Peninsula, collectively called Peninsular Malaysia or West Malaysia. The other two states are on the island of Borneo, and the remaining one federal territory consists of islands offshore of Borneo; these are collectively referred to as East Malaysia or Malaysian Borneo.

The states and federal territories of Malaysia are as follows:

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



1.3.1 Population of Malaysia

Malaysia is a fast-developing country in Southeast Asia with a population of 32.5 million in the third quarter (Q3) of 2018. According to the 2010 census, the population consists of a majority of Malays (50.8%) in addition to other ethnic groups, mainly Chinese (21%), Indian (6%) and other minority groups forming the remaining 11% of the population.

The Malays were the original inhabitants of peninsular Malaysia and together with the indigenous peoples in Sabah and Sarawak (known as “Bumiputera”) form the largest community in Malaysia at 62%. Malaysian Chinese are descendants of migrants from South China, while Malaysian Indians were descended from migrants from Southern India. Aboriginal people known as Orang Asli are present in the east and peninsular Malaysia.

1.4 Data Collection

Data were manually entered into the electronic database on MyTalasemia. The types of data collected are guided by the registry design with data elements grouped into several categories including socio-demographic, clinical characteristics, laboratory test results, types of treatment received, death records and other complications.

The management of registry data requires attention to detail, constant feedback to the participating centre Research Assistants (RA) to complete datasets, and effort to update patients’ new information in a timely manner.

1.5 Participating Centres

1.5.1 Johor

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

1.5.2 Kedah

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

1 Introduction

1.5.3 Kelantan

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

1.5.4 Melaka

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

1.5.5 Negeri Sembilan

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

1.5.6 Pahang

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

1.5.7 Perak

1. Hospital Raja Permaisuri Bainun, Ipoh
2. Hospital Kuala Kangsar
3. Hospital Seri Manjung
4. Hospital Slim River
5. Hospital Taiping
6. Hospital Teluk Intan
7. Hospital Gerik

1.5.8 Perlis

1. Hospital Tuanku Fauziah, Kangar

1.5.9 Pulau Pinang

1. Hospital Pulau Pinang
2. Hospital Seberang Jaya

1.5.10 Sabah

1. Hospital Beaufort
2. Hospital Beluran
3. Hospital Duchess of Kent, Sandakan
4. Hospital Keningau
5. Hospital Kinabatangan
6. Hospital Kota Belud
7. Hospital Kota Marudu

8. Hospital Kuala Penyu
9. Hospital Kudat
10. Hospital Kunak
11. Hospital Lahad Datu
12. Hospital Wanita & Kanak-Kanak Sabah, Likas
13. Hospital Papar
14. Hospital Pitas
15. Hospital Queen Elizabeth, Kota Kinabalu
16. Hospital Ranau
17. Hospital Semporna
18. Hospital Sipitang
19. Hospital Tambunan
20. Hospital Tawau
21. Hospital Tenom

1.5.11 Sarawak

1. Hospital Bintulu
2. Hospital Umum Sarawak, Kuching
3. Hospital Lawas
4. Hospital Limbang
5. Hospital Miri
6. Hospital Sibu
7. Hospital Sarikei
8. Hospital Sri Aman
9. Hospital Betong

1.5.12 Selangor

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

1.5.13 Terengganu

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

1.5.14 Wilayah Persekutuan Kuala Lumpur

1. Institute of Paediatrics, Hospital Kuala Lumpur
2. Pusat Perubatan Universiti Kebangsaan Malaysia, Cheras
3. Pusat Perubatan Universiti Malaya, Petaling Jaya

1.5.15 Wilayah Persekutuan Labuan

1. Hospital Labuan

1.5.16 Wilayah Persekutuan Putrajaya

1. Hospital Putrajaya

2 Registry Report

There are 137 government hospitals including three university hospitals (Pusat Perubatan Universiti Kebangsaan Malaysia (PPUKM), Hospital Universiti Sains Malaysia (HUSM), and Pusat Perubatan Universiti Malaya (PPUM)) in Malaysia. However, only 110 hospitals manage thalassaemia patients. As of 28th November 2018, 8681 thalassaemia patients have been registered in the MTR. The total number of living patients in Malaysia is 7984. Table 2.1 depicts the distribution of thalassaemia patients according to their current status.

Table 2.1: Distribution of Patients in Malaysia by Vital Status

Vital Status	Patients
Alive	7240
Cumulative Reported Cured by Stem Cell Therapy	130
Lost to Follow-Up	614
Total	7984
Cumulative Reported Deaths	697
Total	8681

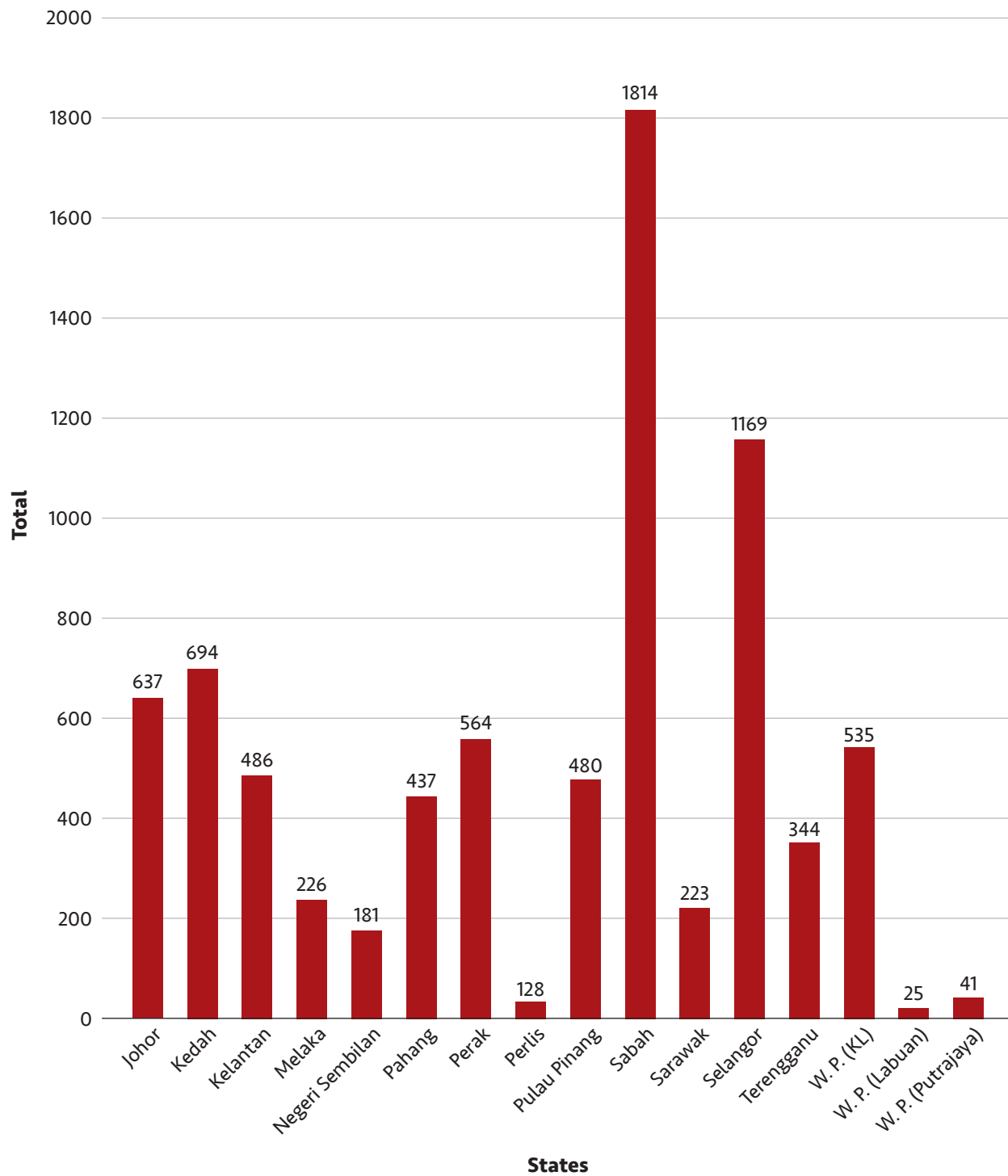
Table 2.2: Number of Thalassaemia Patients by State (2014-2018)

State	2014		2015		2016		2017		2018	
	No.	%	No.	%	No.	%	No.	%	No.	%
Johor	497	7.30	541	7.50	587	7.72	617	7.83	637	7.98
Kedah	649	9.54	668	9.26	680	8.94	688	8.73	694	8.69
Kelantan	401	5.89	423	5.86	449	5.90	474	6.01	486	6.09
Melaka	193	2.84	199	2.76	209	2.75	220	2.79	226	2.83
Negeri Sembilan	168	2.47	172	2.38	180	2.37	180	2.28	181	2.27
Pahang	316	4.64	360	4.99	403	5.30	428	5.43	437	5.47
Perak	493	7.24	519	7.19	550	7.23	563	7.14	564	7.06
Perlis	117	1.72	120	1.66	124	1.63	127	1.61	128	1.60
Pulau Pinang	435	6.39	452	6.26	470	6.18	479	6.08	480	6.01
Sabah	1573	23.12	1671	23.15	1740	22.88	1792	22.74	1814	22.72
Sarawak	176	2.59	194	2.69	207	2.72	219	2.78	223	2.79
Selangor	1019	14.97	1075	14.90	1124	14.78	1160	14.72	1169	14.64
Terengganu	271	3.98	289	4.00	321	4.22	339	4.30	344	4.31
W. P. (KL)	456	6.70	482	6.68	501	6.59	530	6.72	535	6.70
W. P. Labuan	15	0.22	19	0.26	20	0.26	25	0.32	25	0.31
W. P. (Putrajaya)	26	0.38	33	0.46	40	0.53	41	0.52	41	0.51
Total	6805	100.00	7217	100.00	7605	100.00	7882	100.00	7984	100.00

Despite being the third most populous state in Malaysia, Sabah showed the largest number of registered thalassaemia cases in Malaysia, with a total of 1814 patients (22.72%). This is followed by Selangor, Kedah and Johor with 1169, 694 and 637 patients, respectively. Meanwhile, Wilayah Persekutuan Putrajaya and Labuan only have 41 and 25 patients, respectively.

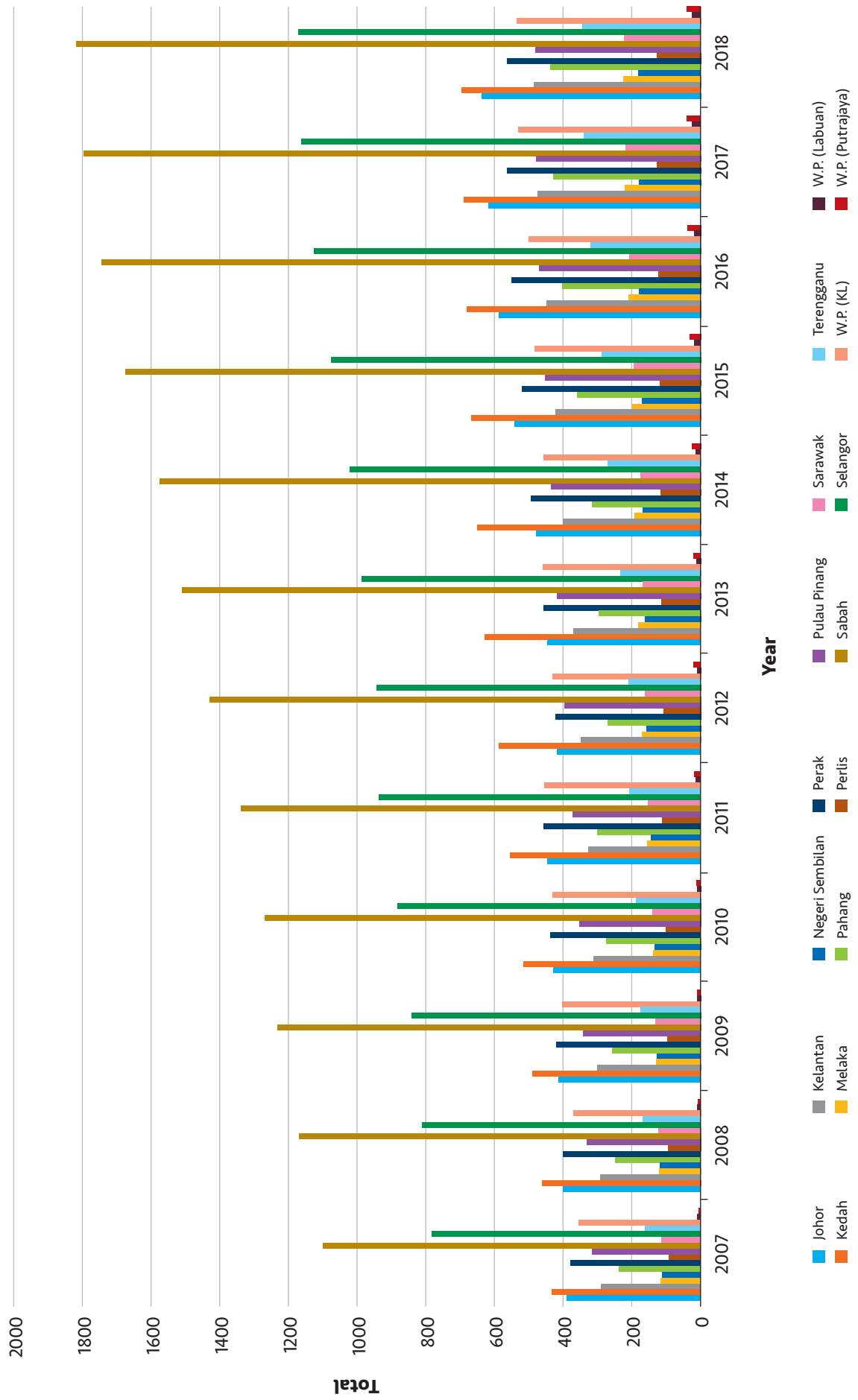


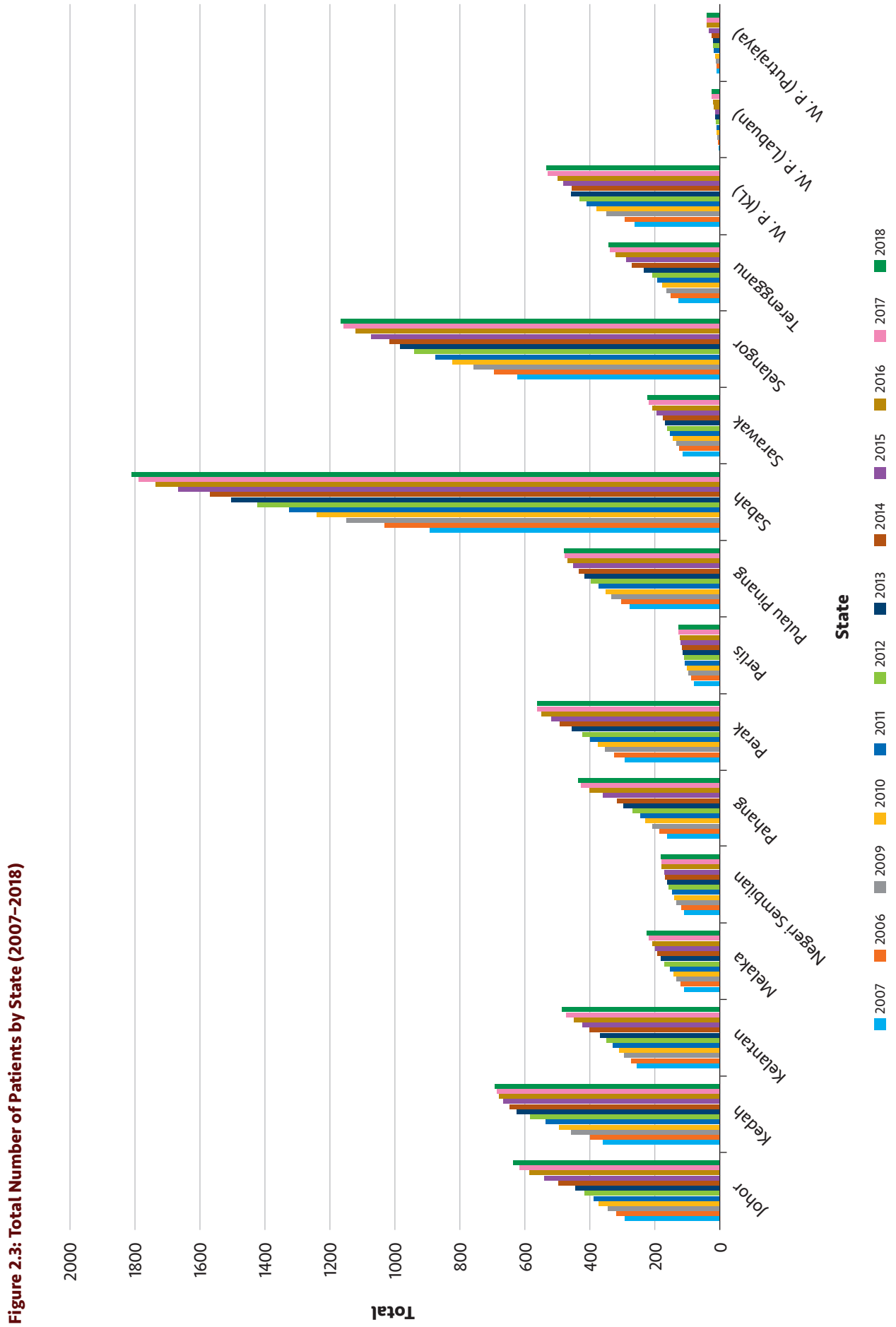
Figure 2.1: Total Number of Thalassaemia Patients by State in 2018



2 Registry Report

Figure 2.2: Total Number of Patients by Year (2007-2018)





2 Registry Report

2.1 Ten Centres with the Highest Number of Patients

Hospital Ampang in Selangor has the highest number of thalassaemia patients among the 110 participating hospitals. It is the national referral centre for adult haematology patients, besides being located in the Klang Valley. The hospital also receives patients mainly from the Federal Territory of Kuala Lumpur.

Table 2.3: Top 10 Centres in Malaysia by Number of Thalassaemia Patients

Centre	No. of Patients (n)
Ampang	703
Sultanah Bahiyah, Alor Setar	370
Queen Elizabeth, Kota Kinabalu	358
Raja Permaisuri Bainun, Ipoh	340
Hospital Wanita & Kanak-Kanak Sabah, Likas	309
Sultanah Aminah, Johor Bahru	300
Pulau Pinang	255
Tengku Ampuan Afzan, Kuantan	250
Seberang Jaya	230
Raja Perempuan Zainab II, Kota Bharu	229

2.2 Patients' Status

The patient's status in our registry can be categorised into either alive, deceased, cured by stem cell transplant, lost to follow-up or transferred to another centre. However, only patients who are alive, lost to follow-up or cured by stem cell transplant were counted as total number of patients in the registry's report. Thus:

- Total number of living patients: 7240
- Cumulative deceased patients up to 2018: 697
- Lost to follow-up: 614
- Cured by stem cell transplant : 130

2.3 Birth Summary

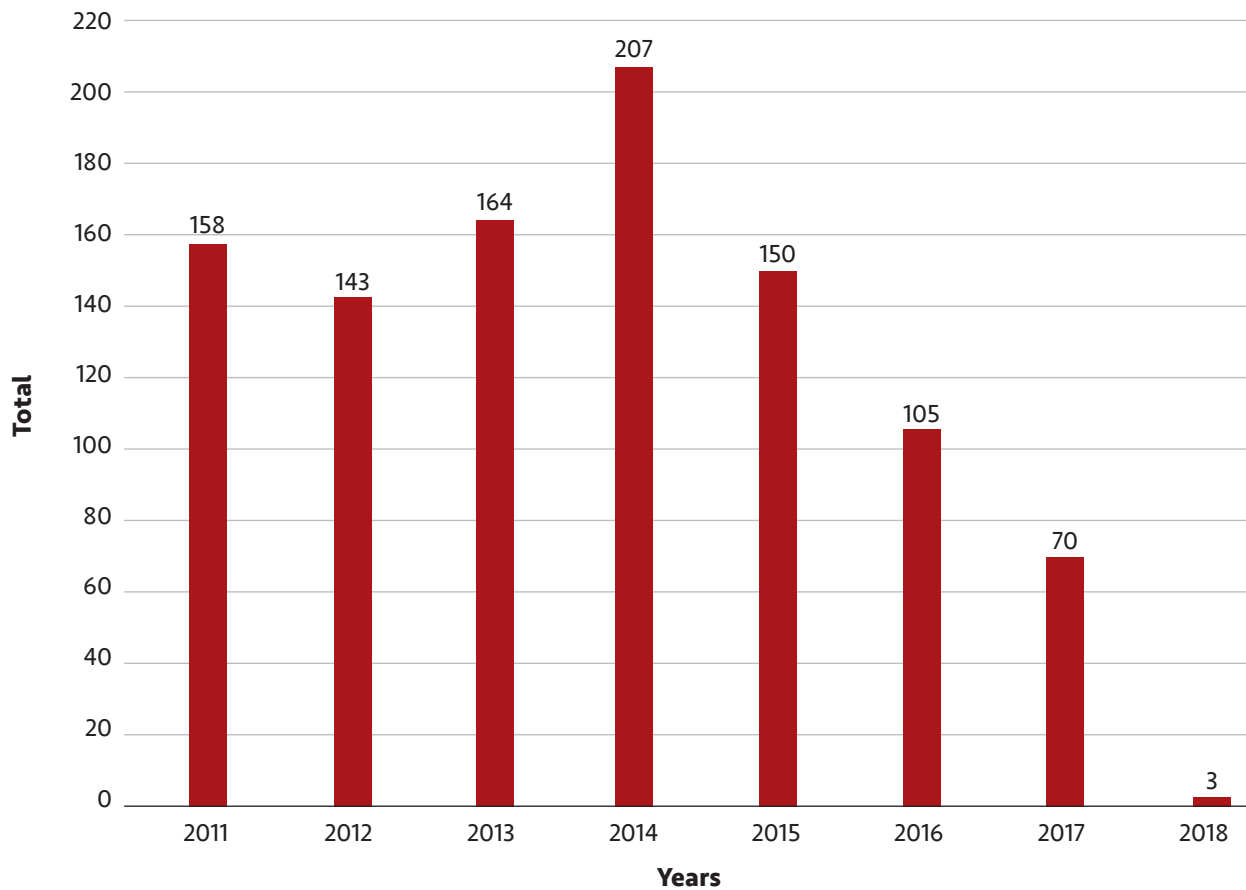
The Ministry of Health of Malaysia estimated that a total of 150-300 babies are born annually with severe thalassaemia syndromes (Ismail *et al.*, 2006). This is in line with data from the registry shown in Table 2.4 for new thalassaemia births from the year 2011 onwards. The number of births was also proportional to the total number of patients in each state of Malaysia. Sabah recorded the highest number of new births each year, followed by Kedah, Selangor and Wilayah Persekutuan Kuala Lumpur.

New births of thalassaemia patients may be registered later than their year of births, depending on the severity of anaemia and age of disease presentation to the hospital. Severe beta thalassaemia major would usually present at 4-6 months of age while the HbE-beta thalassaemia may present much later in the toddler age group. Thalassaemia intermedia or HbH disease patients sometimes present symptoms in their teens or adulthood as an incidental finding. Some are diagnosed when they become pregnant and investigated for anaemia in pregnancy.

Patients with milder forms of thalassaemia syndromes who are asymptomatic may not be diagnosed unless they undergo proper haemoglobin analysis. These group of patients would still need to be followed-up as they require proper genetic counselling and may potentially develop specific issues later in life.

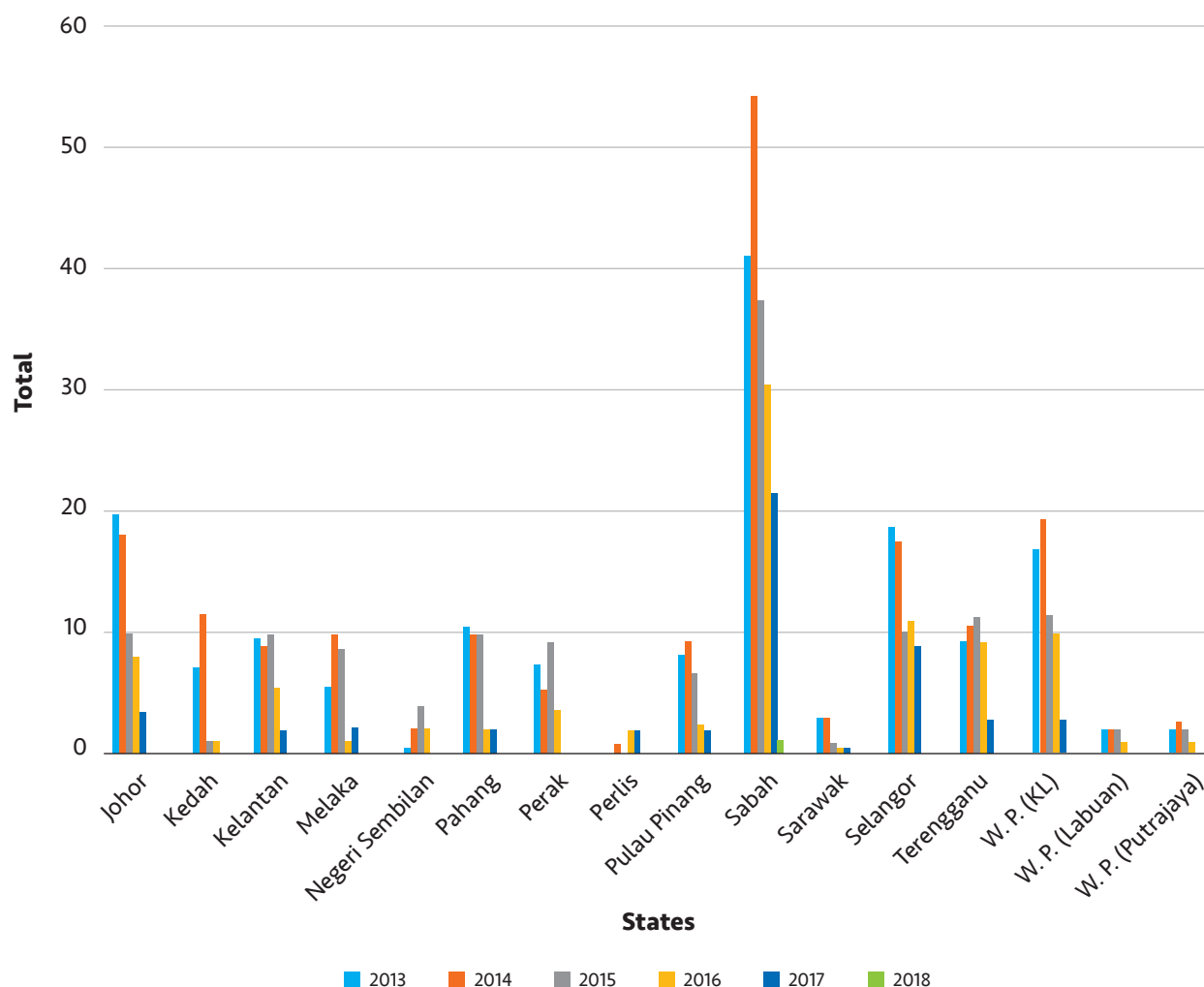
Table 2.4: Number of New Births by State (2011-2018)

State	2011	2012	2013	2014	2015	2016	2017	2018
Johor	6	13	20	18	12	11	7	0
Kedah	23	18	7	13	3	4	2	0
Kelantan	6	5	9	12	12	5	2	1
Melaka	3	8	6	10	8	3	5	0
Negeri Sembilan	3	3	1	2	5	2	0	0
Pahang	6	5	12	11	9	4	2	0
Perak	7	4	7	5	9	3	0	0
Perlis	2	2	0	1	0	2	2	0
Pulau Pinang	6	6	7	10	6	3	2	0
Sabah	48	49	42	60	46	30	23	2
Sarawak	4	3	6	7	2	3	5	0
Selangor	14	14	18	21	10	13	10	0
Terengganu	6	5	9	11	12	9	3	0
W. P. (KL)	21	8	16	19	10	10	7	0
W. P. Labuan	1	0	2	2	2	2	0	0
W. P. (Putrajaya)	2	0	2	5	4	1	0	0
Total	158	143	164	207	150	105	70	3

Figure 2.4: Total Number of New Births (2011-2018)

2 Registry Report

Figure 2.5: Number of New Births by State (2013-2018)



2.4 Death Summary

Morbidity and mortality in thalassaemia patients are the result of chronic transfusion-induced iron overload that may lead to several fatal complications. A total of 697 patients (8.03%) of 8681 registered patients have died since 2007 (Table 2.1). The data demonstrated that heart disease remains the leading cause of death (Table 2.6).

The cumulative reported number of deaths by state is presented in Table 2.5. Sabah has the highest number of deaths followed by Kuala Lumpur, Selangor and Perak. Negeri Sembilan, Melaka, Putrajaya and Labuan had recorded fewer number of deaths.

Out of the 697 deaths recorded, 89 had incomplete data. The leading cause of death among the 608 thalassaemia patients with verifiable data in Malaysia were cardiac-related/cardiac failure (254 cases, 41.78%) and infections (232 cases, 38.16%). Other specified causes were motor vehicle accidents (18 cases, 2.96%), liver diseases (16, 2.63%), tumour and malignancies (15, 2.47%), endocrine complications (13, 2.14%), multiple organ dysfunction (7, 1.15%), surgical complications (7, 1.15%), thrombosis (6, 0.99%), CNS events (5, 0.82%) and other causes (35, 5.76%). The case records for deaths with incomplete data were unavailable and may have been archived, leaving the details unrecorded into the registry.

Table 2.5: Cumulative Reported Number of Deaths by State until December 2018

State	No. of Patients
Johor	30
Kedah	31
Kelantan	29
Melaka	2
Negeri Sembilan	6
Pahang	36
Perak	44
Perlis	12
Pulau Pinang	12
Sabah	303
Sarawak	21
Selangor	64
Terengganu	23
W. P. (KL)	79
W. P. Labuan	4
W. P. (Putrajaya)	1
Total	697

The data is similar to the reported studies in other countries, in which the major cause of death is cardiac complication or dysfunction (33.14%). Studies suggest that heart failure in these patients may due to constrictive pericarditis and cardiomyopathy secondary to iron overload (Borgna-Pinatti *et al.*, 2004; Ladis *et al.*, 2005; Fucharoen *et al.*, 2000).

It is of interest that the second-leading cause of death (38.16%) was infections. Infections have also been reported as the main cause of death in beta thalassaemia patients (Sakran *et al.*, 2012; Chirico *et al.*, 2015). Iron accumulation in the liver may cause fibrosis and cirrhosis, and several complications such as endocrine abnormalities and multiple organ dysfunctions. Other causes include motor vehicle accident, drowning, falls, tumours, thrombosis and CNS events.

Table 2.6: Cumulative Causes of Death Since 2007

State	No. of Patients	Percentage (%)
Cardiac-Related/Cardiac Failure	254	41.78
Infections	232	38.16
Motor Vehicle Accidents	18	2.96
Liver Disease	16	2.63
Tumours & Malignancies	15	2.47
Endocrine Complications	13	2.14
Surgical Complications	7	1.15
Multiple Dysfunctional Organ	7	1.15
Thrombosis	6	0.99
Central Nervous System Event	5	0.82
Others	35	5.76
Total	608	100

2 Registry Report

Table 2.7: Number of Thalassaemia Patient Deaths by State (2008-2018)

State	2008	2009	2010	2011	2012	2013	2014	2015	2016	2017	2018
Johor	1	6	4	1	2	5	0	2	1	4	4
Kedah	0	7	2	1	5	1	3	1	6	2	3
Kelantan	1	2	5	1	2	1	1	4	1	0	4
Melaka	0	0	0	0	0	0	0	0	0	0	1
Negeri Sembilan	1	0	2	1	0	1	0	0	0	0	0
Pahang	0	6	3	3	1	2	1	1	4	6	3
Perak	2	4	3	2	6	1	1	0	4	4	5
Perlis	0	2	2	1	0	2	2	0	0	0	0
Pulau Pinang	3	1	0	1	0	0	0	3	0	1	1
Sabah	28	28	26	21	26	30	18	16	16	21	18
Sarawak	0	1	1	2	1	1	1	2	3	0	3
Selangor	5	4	6	6	5	5	4	7	5	6	9
Terengganu	3	1	2	1	3	0	4	4	2	0	1
W. P. (KL)	3	6	2	2	1	4	1	6	1	0	1
W. P. Labuan	0	2	1	0	0	0	0	0	1	0	0
W. P. (Putrajaya)	0	0	1	0	0	0	0	0	0	0	0
Total	47	70	60	43	52	53	36	46	44	44	53

Figure 2.6: Total Number of Deaths by Year (2008-2018)

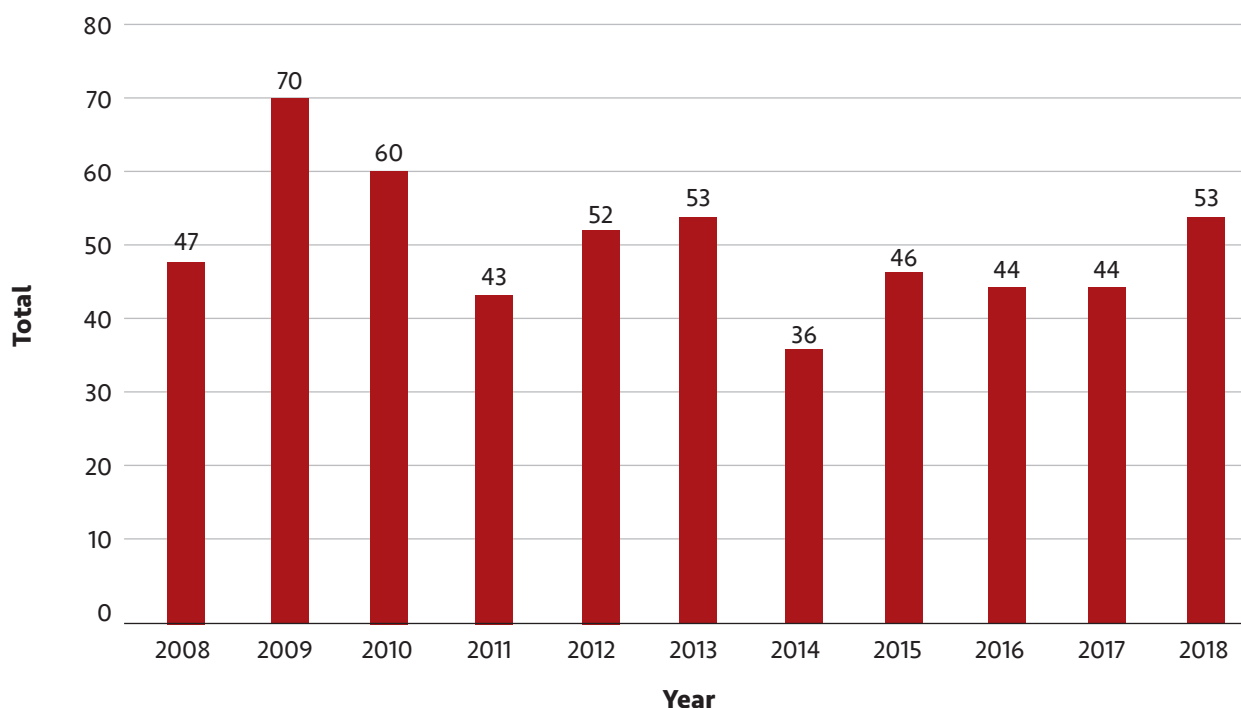
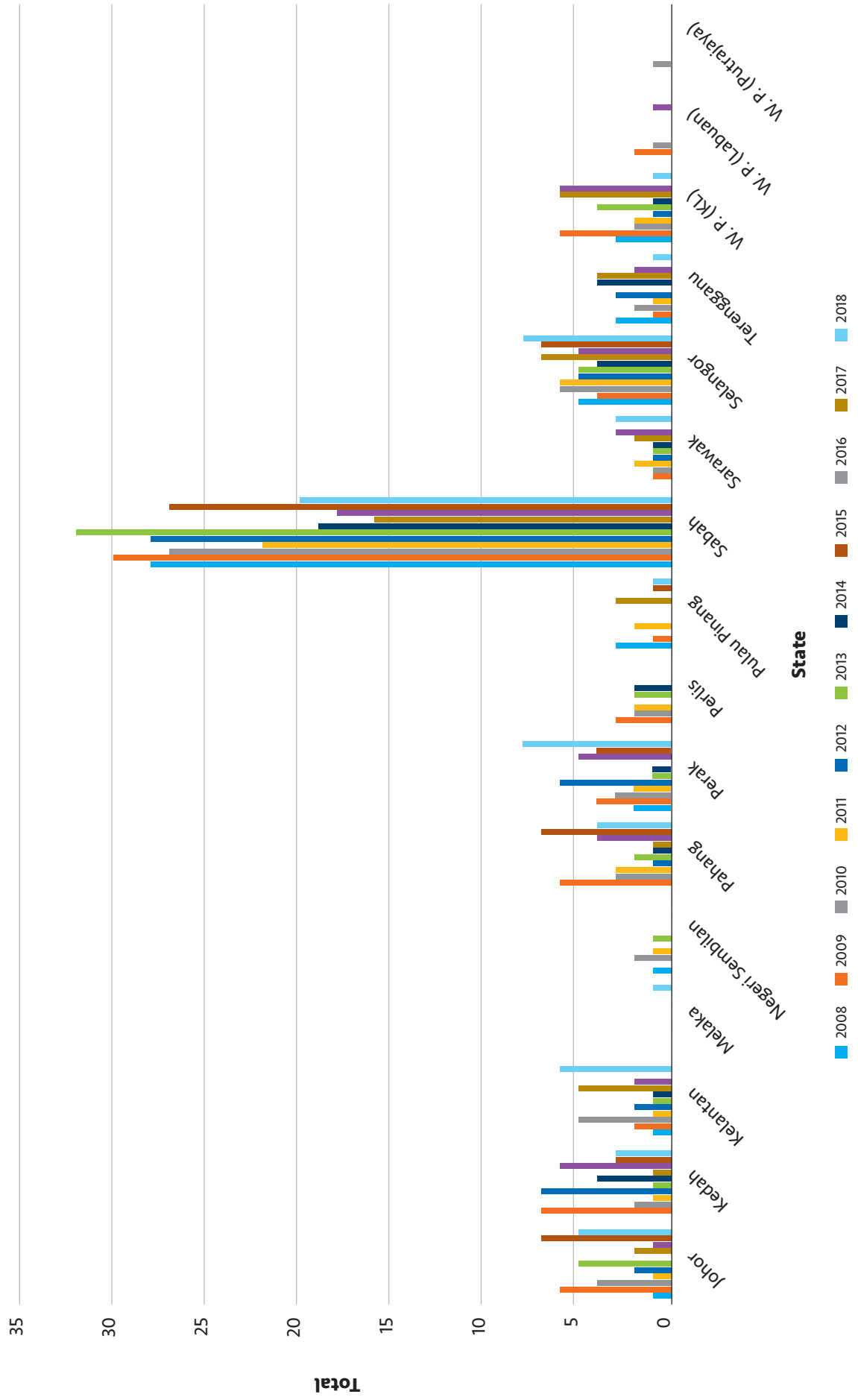


Figure 2.6 demonstrates the cumulative reported thalassaemia deaths over the years. In total, 694 thalassaemia patients have died. The highest number of deaths was recorded in 2009 (70 cases).

Figure 2.7: Total Number of Deaths by State (2008-2018)



2 Registry Report

2.5 Patient Demographics

Healthcare registers have been defined as organised systems with uniform data aimed at comprehensive coverage of a target population with a particular disease, condition or exposure. Thus, it is essential to establish a national thalassaemia registry collecting specific patient information into the registry database. The thalassaemia registry collects a set of patients' socio-demographic details, such as centre of treatment, name, date of birth, home address, contact phone number, gender, ethnicity, and sibling rank.

Though the core element in the information system is the diagnosis and other clinical characteristics, the demographic characteristics also play a major role for public health surveillance, healthcare monitoring and administrative purposes for specific regions or overall national data.

Specific MTR forms have been developed over the years to manage the data collection before data entry in the web system. Reported errors or modifications are managed centrally to prevent any double entries in the registry.

2.5.1 Age

Out of 7984 thalassaemia patients in Malaysia, the age group between 11-15 years old have the highest number of patients with 1394 patients (17.46%), followed by 16-20 years old and 6-10 years old with 1286 (16.11%) and 1272 (15.93%) patients, respectively. The data also indicate a smaller number of the patients above 50 years old (278 patients, 3.48 %).

Table 2.8: Distribution of Thalassaemia Patients According to Age Group by State

State	Total No. of Patients	Age Group (years)																	
		0-4.9	5-9.9	10-14.9	15-19.9	20-24.9	25-29.9	30-34.9	35-39.9	40-44.9	45-49.9	50-54.9	55-59.9	60-64.9	65-69.9	70-74.9	75-79.9	80-84.9	85-89.9
Johor	637	48	110	101	108	97	59	32	30	15	16	7	8	3	3	0	0	0	0
Kedah	694	22	130	154	145	122	64	28	7	8	7	3	1	1	1	0	0	0	0
Kelantan	486	32	70	82	79	89	57	25	13	9	15	6	3	2	1	0	1	2	0
Melaka	226	26	45	48	29	48	22	7	0	1	0	0	0	0	0	0	0	0	0
Negeri Sembilan	181	9	30	18	37	27	19	13	7	7	5	3	3	3	0	0	0	0	0
Pahang	437	26	70	62	61	61	57	35	22	13	12	5	8	1	1	2	0	0	1
Perak	564	17	59	87	81	91	64	56	35	21	16	11	11	10	2	2	1	0	0
Perlis	128	5	12	24	25	35	16	6	1	0	1	0	1	2	0	0	0	0	0
Pulau Pinang	480	21	55	74	78	68	51	35	30	24	15	13	6	1	4	1	3	0	1
Sabah	1814	161	370	392	327	245	138	80	41	17	12	11	1	6	10	2	1	0	0
Sarawak	223	17	34	35	33	42	16	21	9	2	6	4	1	1	0	1	1	0	0
Selangor	1169	54	119	124	113	165	139	142	104	84	42	25	17	17	13	4	3	1	3
Terengganu	344	35	53	68	61	30	31	25	15	7	4	9	1	4	0	0	1	0	0
W. P. (KL)	535	46	95	106	104	71	57	40	13	1	0	1	0	0	1	0	0	0	0
W. P. Labuan	25	6	9	5	3	1	0	0	0	0	1	0	0	0	0	0	0	0	0
W. P. (Putrajaya)	41	10	11	14	2	2	0	0	0	2	0	0	0	0	0	0	0	0	0
Total	7984	535	1272	1394	1286	1194	790	545	327	211	152	98	61	51	36	13	11	3	5

Figure 2.8: Total Number of Patients by Age Group

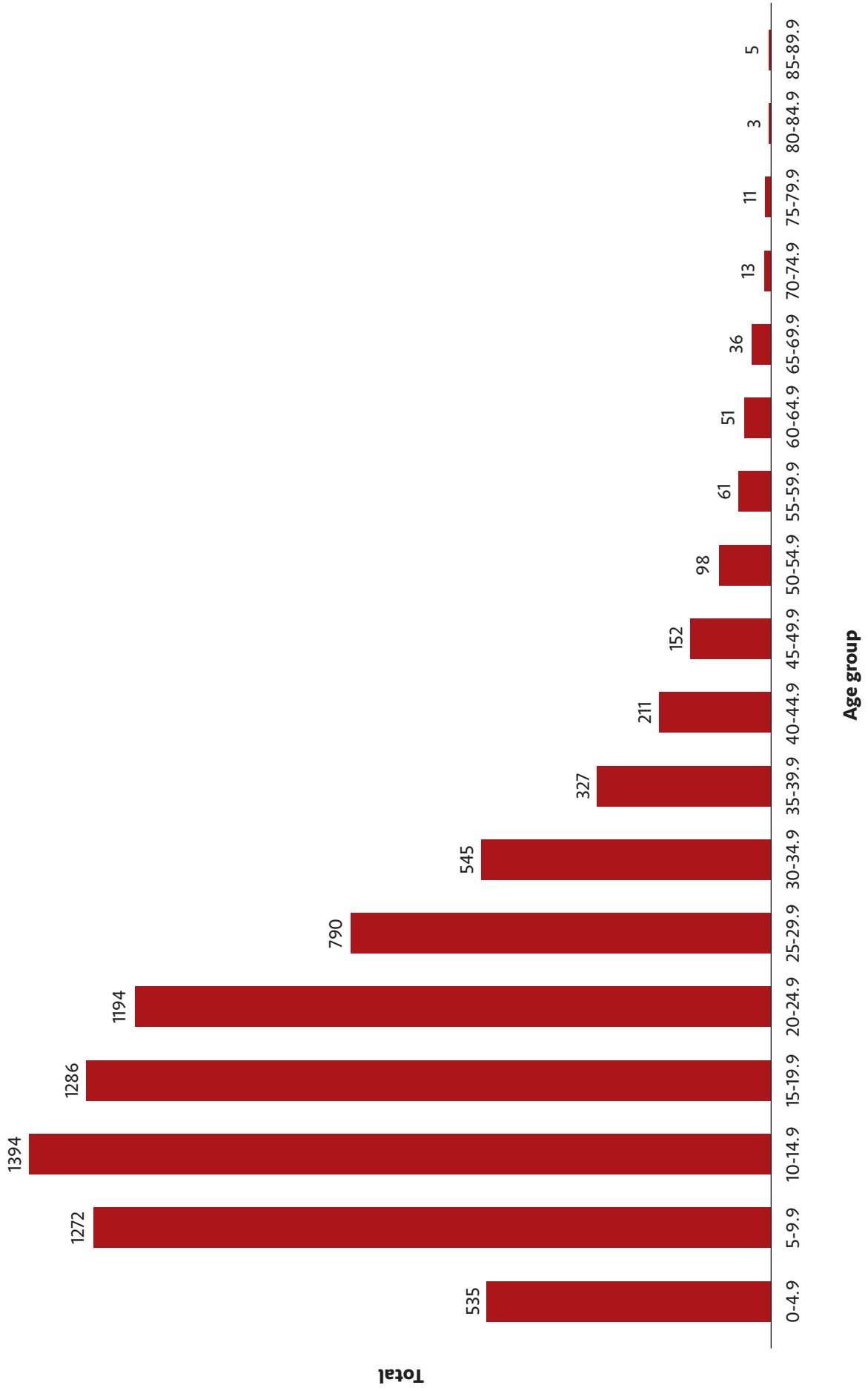
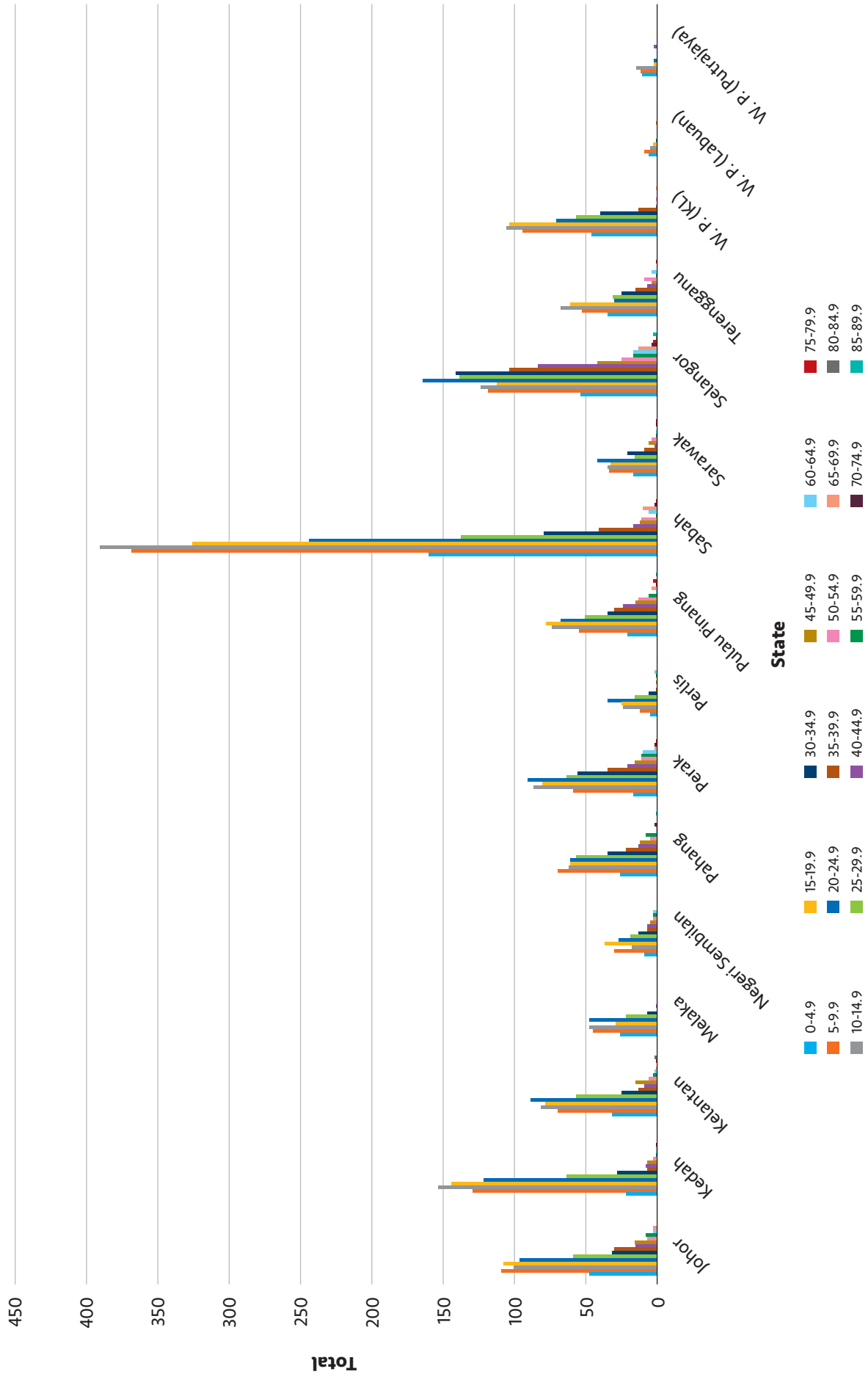


Figure 2.9: Distribution of Patients According to Age Group by State



2.5.2 Gender

Of the 7984 thalassaemia patients in Malaysia, 3960 patients (49.60%) are male and 4024 patients (50.40%) are female. Female patients are slightly more in number than male patients, although the difference is small. Other reports in several countries demonstrate more male patients than female patients.

Figure 2.10: Distribution of Patients by Gender

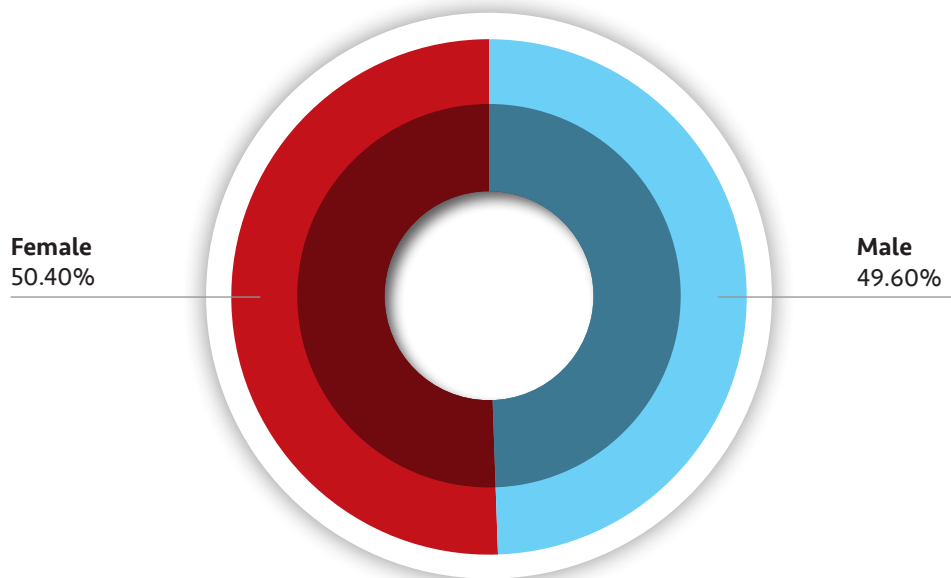
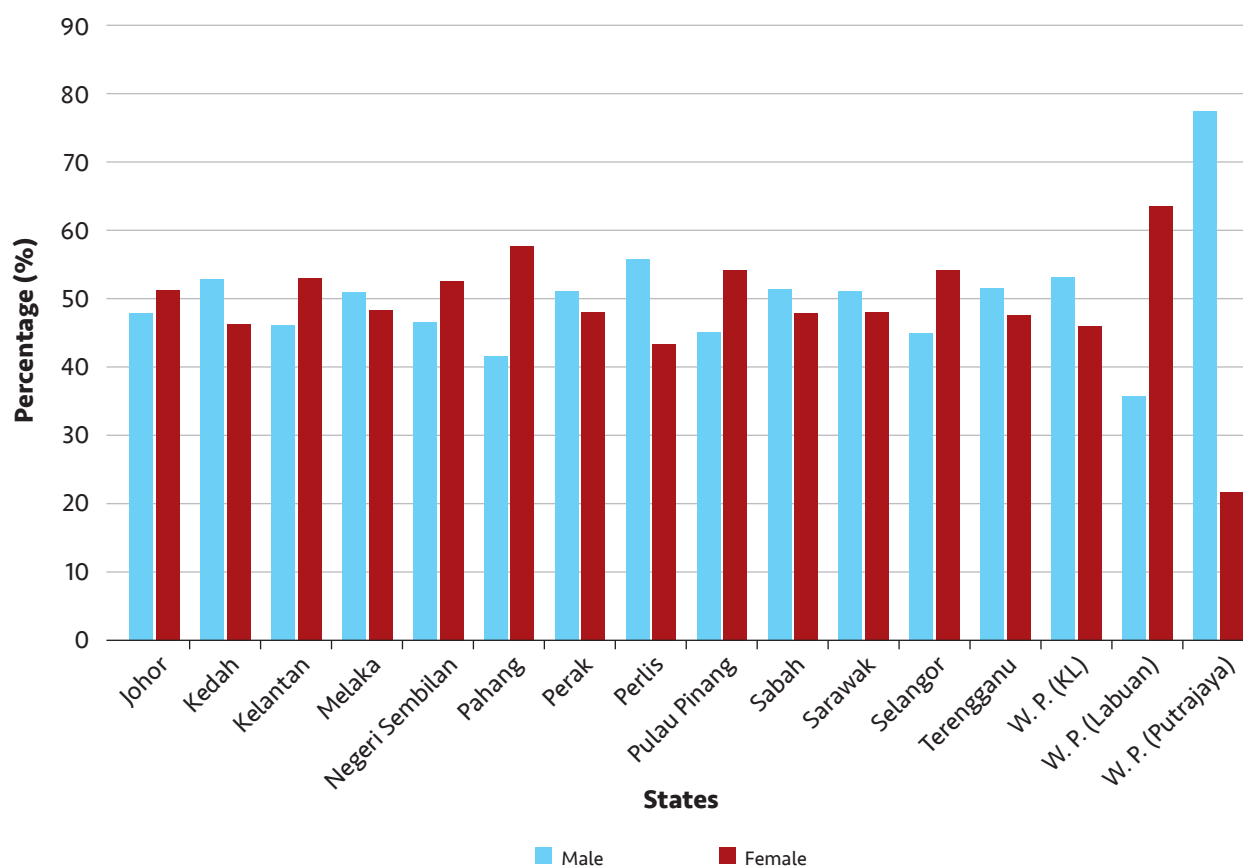


Table 2.9: Distribution of Patients According to Gender by State

State	Total	Male		Female	
		No	%	No	%
Johor	637	308	48.35	329	51.65
Kedah	694	370	53.31	324	46.69
Kelantan	486	226	46.50	260	53.50
Melaka	226	116	51.33	110	48.67
Negeri Sembilan	181	85	46.96	96	53.04
Pahang	437	183	41.88	254	58.12
Perak	564	291	51.60	273	48.40
Perlis	128	72	56.25	56	43.75
Pulau Pinang	480	218	45.42	262	54.58
Sabah	1814	939	51.76	875	48.24
Sarawak	223	115	51.57	108	48.43
Selangor	1169	530	45.34	639	54.66
Terengganu	344	179	52.03	165	47.97
WP (KL)	535	287	53.64	248	46.36
WP (Labuan)	25	9	36.00	16	64.00
WP (Putrajaya)	41	32	78.05	9	21.95
Total	7984	3960	49.60	4024	50.40

2 Registry Report

Figure 2.11: Distribution of Patients According to Gender by State



2.5.3 Ethnic group

The three major ethnic groups in Malaysia comprise the Malays, Chinese and Indian; however, in the Borneo states of Sabah and Sarawak, there exists multiple sub-ethnic groups including the Kadazan-Dusun which is the largest indigenous ethnic group in Sabah. Population studies have 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).

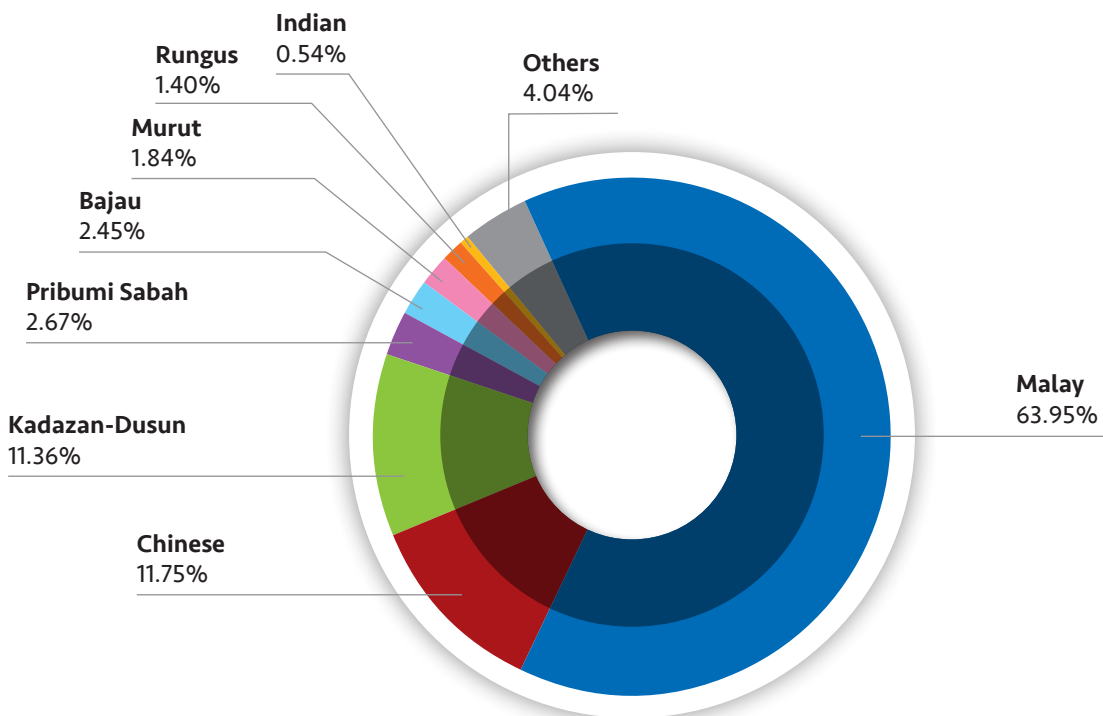
The molecular defects producing beta thalassaemia are heterogeneous, and each ethnic group possesses its own specific set of mutations according to several previous studies. Alpha thalassaemia has also been reported and its fatal condition, Hb Bart's hydrops fetalis, is present mainly in the Malaysian Chinese. The Malays possess the single alpha globin gene deletion that produces an asymptomatic disorder and thalassaemia in Malaysian Indians are only rarely observed.

Table 2.10 shows that Malay patients formed the largest group in Malaysia with 5106 patients (63.95%). This is followed by the Chinese with 938 patients (11.75%), Kadazan-Dusun with 907 patients (11.36%), Pribumi Sabah with 213 patients (2.67%), Bajau with 196 patients (2.45%), Murut with 147 patients (1.84%) and Indian with 43 patients (0.54%). The remaining 322 patients (4.03%) patients are either of Orang Asli, Thai, Foreigner, Iban, Pribumi Sarawak, Sino-Kadazan, Kedayan, Bidayuh, Melanau, Mixed or Kedayan ethnicity. The "Others" ethnicity status may refer to other ethnic groups that are not specified.

Table 2.10: Distribution of Patients by Ethnic Group

Ethnic	No	Percentage (%)
Malay	5106	63.95
Chinese	938	11.75
Kadazan-Dusun	907	11.36
Pribumi Sabah	213	2.67
Bajau	196	2.45
Murut	147	1.84
Rungus	112	1.40
Indian	43	0.54
Others:		
Orang Asli	43	0.54
Thai	44	0.55
Foreigner	68	0.85
Iban	20	0.25
Pribumi Sarawak	17	0.21
Sino-Kadazan	27	0.34
Bidayuh	7	0.09
Mixed	34	0.43
Melanau	4	0.05
Others	58	0.73
Total	7984	100.00

Figure 2.12: Distribution of Patients by Ethnic Group



2 Registry Report

Table 2.11: Distribution of Patients According to Major Ethnic Groups by State

State	Total No. of Patients	Malay	Chinese	Kadazan-Dusun	Bajau	Pribumi Sabah	Murut	Rungus	Indian	Others
Johor	637	483	114	15	0	5	2	0	11	8
Kedah	694	654	17	0	0	0	0	0	2	21
Kelantan	486	461	14	0	0	0	0	0	0	11
Melaka	226	178	40	0	1	1	0	0	3	3
Negeri Sembilan	181	141	29	1	0	0	0	0	3	7
Pahang	437	393	31	0	1	0	0	0	0	12
Perak	564	421	113	1	0	1	0	0	9	19
Perlis	128	122	3	0	0	0	1	0	0	2
Pulau Pinang	480	379	84	3	0	0	0	0	4	10
Sabah	1814	101	96	858	185	202	139	111	1	123
Sarawak	223	95	86	5	0	0	0	0	0	34
Selangor	1169	902	190	16	4	1	3	1	7	45
Terengganu	344	337	6	0	0	0	0	0	0	1
WP (Kl)	535	393	113	2	0	0	1	0	3	23
WP (Labuan)	25	6	1	6	5	3	1	0	0	3
WP (Putrajaya)	41	40	1	0	0	0	0	0	0	0
Total	7984	5106	938	907	196	213	147	112	43	322

Sabah has as the highest total number of patients (1814, 22.72%), dominated by the Kadazan-Dusun which is the major ethnic group in Sabah. The Kadazan-Dusun forms the largest group of patients in Sabah, with 858 patients (47.30%).

An analysis based on regional division of Peninsular Malaysia, Sabah and Sarawak has shown a different pattern of distribution of patients by ethnic group. Wilayah Persekutuan Kuala Lumpur and Putrajaya were grouped into Peninsular Malaysia whereby Wilayah Persekutuan Labuan was grouped into Sabah based on geographical proximity.

Table 2.12: Distribution of Patients According to Major Ethnic Group Based on Geographical Region

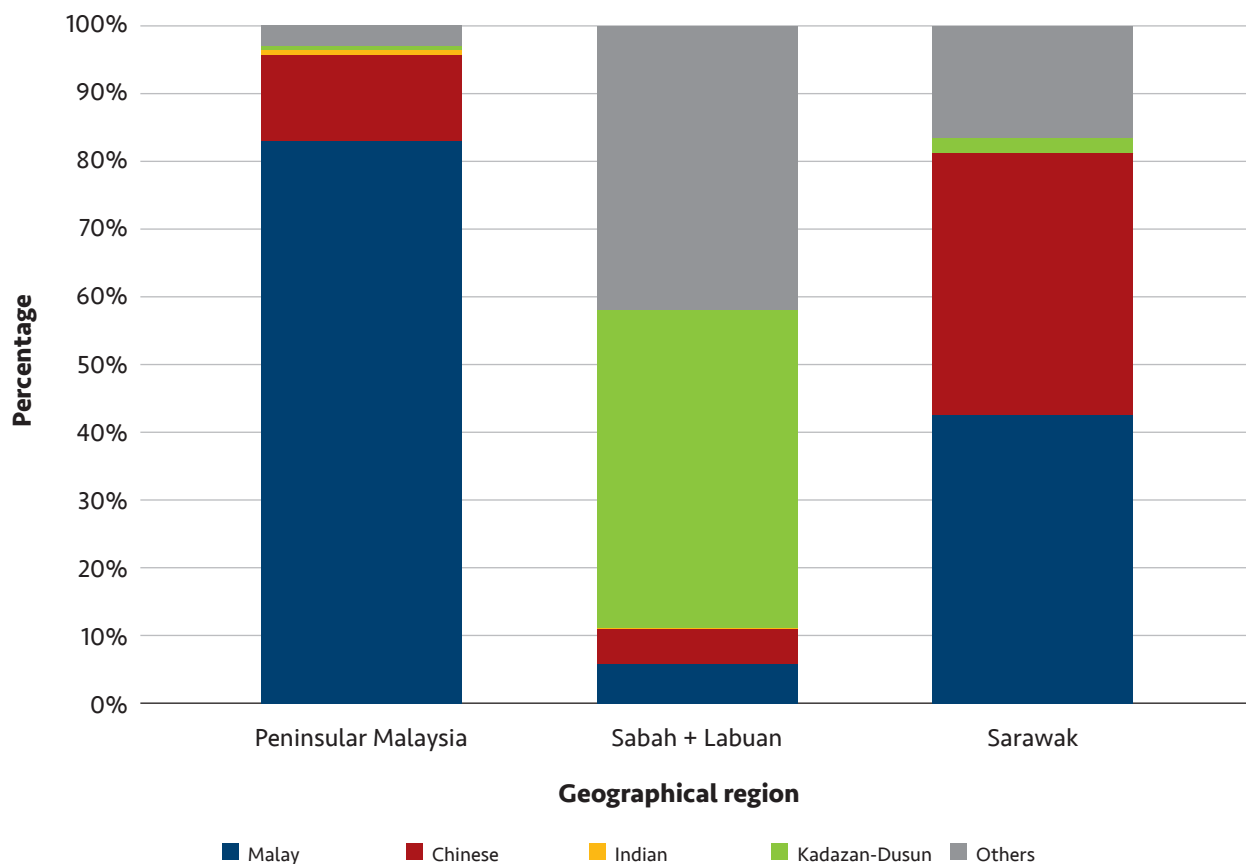
State	Total No. of Patients	Malay		Chinese		Indian		Kadazan-Dusun		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
Peninsular Malaysia	5922	4904	82.81	755	12.75	42	0.71	38	0.64	183	3.09
Sabah + Labuan	1839	107	5.82	97	5.27	1	0.05	864	46.98	770	41.87
Sarawak	223	95	42.60	86	38.57	0	0.00	5	2.24	37	16.59
Total	7984	5106	63.95	938	11.75	43	0.54	907	11.36	990	12.40

Note: % is a calculated against regional total, i.e. not national total.

With the modified regional grouping, the following observations can be made:

- Malay patients form 4904 patients out of 5922 patients (82.81%) in Peninsular Malaysia.
- Kadazan-Dusun patients form 864 out of 1839 patients (46.98%) in Sabah and Wilayah Persekutuan Labuan.
- Malay and Chinese patients form 42.60% and 38.57% of patients in Sarawak, respectively.

Figure 2.13: Distribution of Patients According to Major Ethnic Groups by Geographical Region



2 Registry Report

2.6 Diagnosis

The type of thalassaemias in our registry generally are classified as beta thalassaemia major, beta thalassaemia intermedia, HbE-beta thalassaemia and HbH disease. Others thalassaemia diagnoses includes other forms of thalassaemia, such as HbH Constant Spring, alpha thalassaemia, Hb Adana, HbAE Bart's Constant Spring, heterogenic Hb Lepore Hollandia, delta-beta thalassaemia and other thalassaemia disorders.

The beta thalassaemia syndromes are much more diverse than the alpha thalassaemia syndromes due to the diversity of the mutations that produce the defects in the beta globin gene. Patients who were diagnosed with thalassaemia intermedia have a homozygous or heterozygous beta globin mutation that causes a decrease in beta chain production, but not to the degree that chronic transfusion therapy is required. The terms thalassaemia minor, thalassaemia intermedia and thalassaemia major describe the severity of the symptoms. Thalassaemia minor does not cause any symptoms, whereas thalassaemia major is the most severe.

Table 2.13: Distribution of Patients by Diagnosis in 2018

Diagnosis	No. of Patients	Percentage (%)
Beta Thalassaemia Major	2676	33.52
Beta Thalassaemia Intermedia	748	9.37
HbE-Beta Thalassaemia	2744	34.37
HbH Disease	1458	18.26
Others	358	4.48
Total	7984	100.00

The interaction of HbE with beta thalassaemia results in HbE-beta thalassaemia, an extremely heterogeneous clinical condition. HbE-beta thalassaemia is the most common form of beta thalassaemia in Southeast Asia. As depicted in Table 2.13, the HbE-beta thalassaemia forms the largest group of thalassaemia patients in Malaysia with 2744 patients (34.37%), followed by the beta thalassaemia major with 2676 patients (33.52%), HbH disease with 1458 patients (18.26%), beta thalassaemia intermedia with 748 patients (9.37%), while the remaining 358 patients (4.48%) have others forms of haemoglobinopathies.

Table 2.14: Distribution of Patients According to Diagnosis by Year

Year	Total No. of Patients (n)	Thalassaemia Major		Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
2018	7984	2676	33.52	748	9.37	2744	34.37	1458	18.26	358	4.48
2017	7882	2646	33.57	741	9.40	2700	34.26	1430	18.14	365	4.63
2016	7605	2580	33.93	714	9.39	2594	34.11	1375	18.08	342	4.50
2015	7217	2490	34.50	691	9.57	2440	33.81	1288	17.85	308	4.27
2014	6805	2378	34.94	648	9.52	2313	33.99	1193	17.53	273	4.01
2013	6386	2277	35.66	614	9.61	2169	33.96	1086	17.01	240	3.76
2012	5973	2173	36.38	577	9.66	2031	34.00	985	16.49	207	3.47
2011	5547	2061	37.16	533	9.61	1879	33.87	893	16.10	181	3.26
2010	5164	1970	38.15	498	9.64	1743	33.75	797	15.43	156	3.02
2009	4786	1851	38.68	455	9.43	1607	33.24	598	12.29	275	5.65
2008	4318	1681	38.93	417	9.57	1464	33.52	531	12.07	225	5.11
2007	3859	1511	39.16	367	9.42	1322	33.82	466	11.82	193	4.88

Table 2.14 indicates that number of patients by diagnosis is increasing each year.

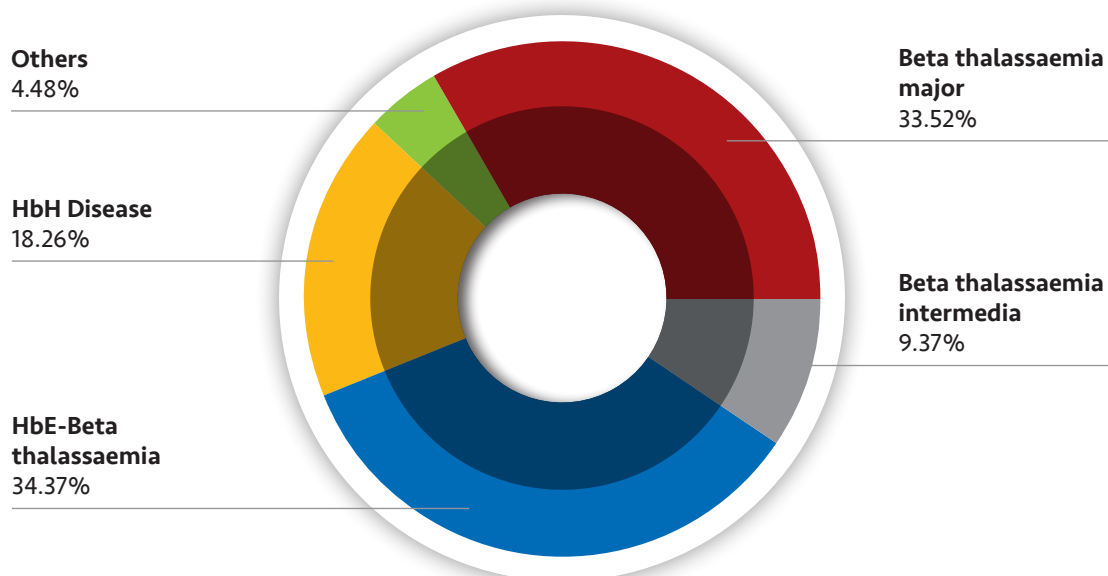
Table 2.15: Distribution of Patients According to Diagnosis by State

State	Total No. of Patients (n)	Thalassaemia Major		Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
		No.	%*	No.	%*	No.	%*	No.	%*	No.	%*
Johor	637	210	32.97	53	8.32	264	41.44	106	16.64	4	0.63
Kedah	694	107	15.42	50	7.20	290	41.79	188	27.09	59	8.50
Kelantan	486	70	14.40	31	6.38	241	49.59	117	24.07	27	5.56
Melaka	226	50	22.12	7	3.10	98	43.36	55	24.34	16	7.08
N. Sembilan	181	51	28.18	21	11.60	67	37.02	27	14.92	15	8.29
Pahang	437	79	18.08	32	7.32	182	41.65	103	23.57	41	9.38
Perak	564	114	20.21	51	9.04	234	41.49	115	20.39	50	8.87
Perlis	128	26	20.31	17	13.28	53	41.41	24	18.75	8	6.25
Pulau Pinang	480	96	20.63	34	7.08	202	42.08	121	25.21	24	5.00
Sabah	1814	1327	73.15	276	15.21	115	6.34	92	5.07	4	0.22
Sarawak	223	94	42.15	23	10.31	42	18.83	56	25.11	8	3.59
Selangor	1169	209	17.88	89	7.61	518	44.31	289	24.72	64	5.47
Terengganu	344	63	18.31	31	9.01	180	52.33	61	17.73	9	2.62
WP (KL)	535	152	28.41	20	3.74	237	44.30	100	18.69	26	4.86
WP (Labuan)	25	15	60.00	6	24.00	0	0.00	1	4.00	3	12.00
WP (Putrajaya)	41	10	24.39	7	17.07	21	51.22	3	7.32	0	0.00
Total	7984										

*Percentage are proportional to each total state respectively.

HbE-beta thalassaemia is common in Peninsular Malaysia (dominated by Malay patients). Table 2.15 shows that most states in Peninsular Malaysia have more than 40% of its total thalassaemia patients diagnosed with HbE-beta thalassaemia. For the indigenous people of East Malaysia, the most common cause for transfusion-dependent beta thalassaemia major is a homozygous state of the Filipino deletion and not HbE-beta thalassaemia.

Figure 2.14: Distribution of Patients by Diagnosis



2 Registry Report

Figure 2.15: Distribution of Patients According to Diagnosis by State

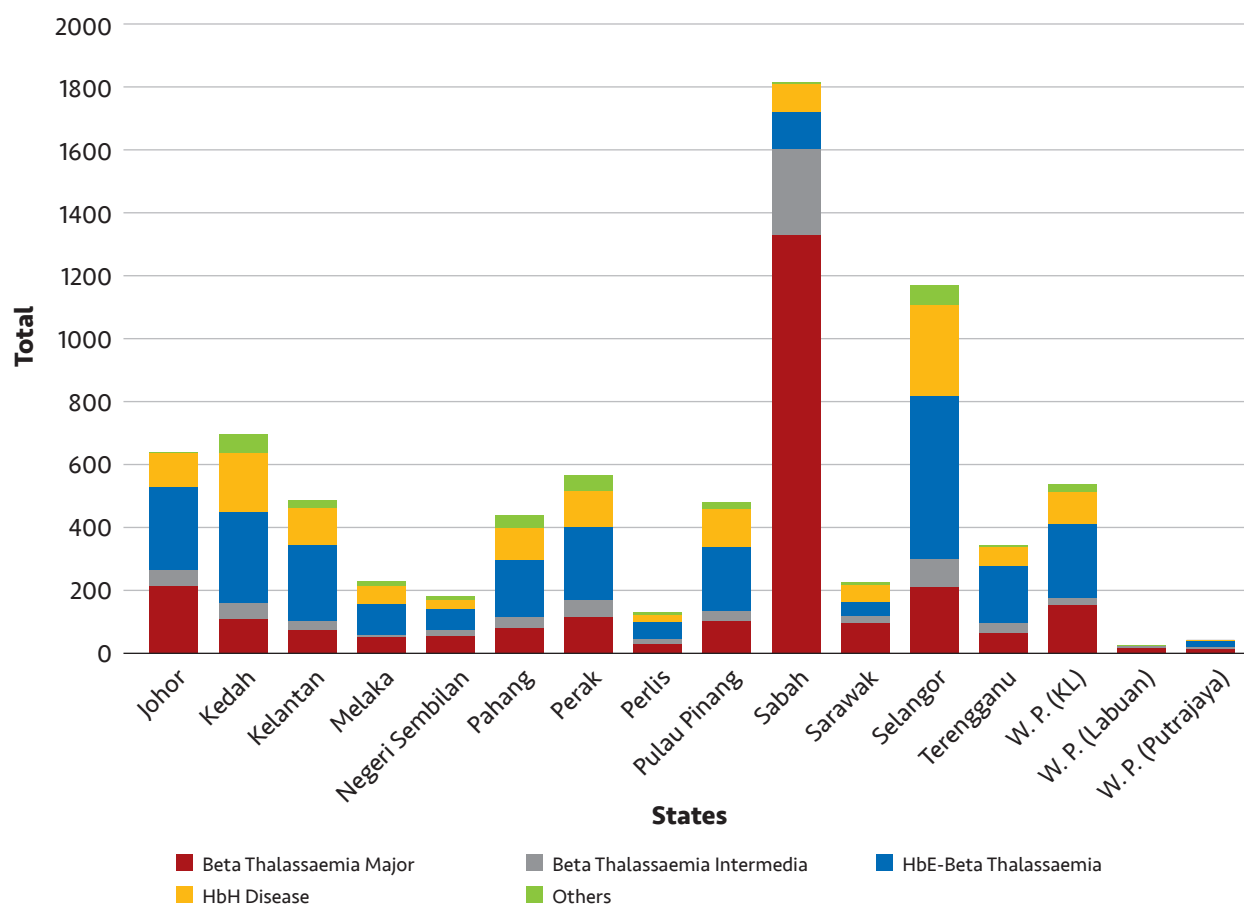


Table 2.16: Distribution of Patients According to Diagnosis by Age Group

Age Group (Years)	Total No. of Patients	Thalassaemia Major		Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
		No.	%	No.	%	No.	%	No.	%	No.	%
0-4.9	535	226	42.24	44	8.22	160	2	77	0.95	28	0.36
5-9.9	1272	463	36.40	105	8.25	399	5	221	2.72	84	1.1
10-14.9	1394	497	35.65	136	9.76	476	5.96	225	2.77	60	0.8
15-19.9	1286	466	36.24	99	7.70	485	6.07	195	2.44	41	0.51
20-24.9	1194	483	40.45	80	6.70	394	4.93	205	2.57	32	0.4
25-29.9	790	268	33.92	64	8.10	310	3.88	132	1.65	16	0.2
30-34.9	545	167	30.64	58	10.64	198	2.48	99	1.23	23	0.3
35-39.9	327	65	19.88	49	14.98	118	1.48	68	0.85	27	0.34
40-44.9	211	19	9.00	30	14.22	81	1.01	66	0.83	15	0.19
45-49.9	152	14	9.21	32	21.05	46	0.58	52	0.65	8	0.1
50-54.9	98	3	3.06	23	23.47	32	0.4	32	0.4	8	0.1
55-59.9	61	2	3.28	11	18.03	20	0.25	22	0.28	6	0.08
60-64.9	51	2	3.92	6	11.76	14	0.18	23	0.29	6	0.08
Above 65	68	1	1.47	11	16.18	11	0.14	41	0.51	4	0.05
Total	7984	2676	33.52	748	9.37	2744	34.37	1458	18.14	358	4.61

In Table 2.16, the highest number of patients are within age range of 5-29.9 years. Out of 2676 of patients with beta thalassaemia major, 2177 patients (81.35%) are between 5-29.9 years of age. Similarly for HbE-beta thalassaemia and HbH disease, 2064 out of 2744 patients (75.22%) and 978 out of 1458 patients (67.08%), respectively, are within the 5-29.9 years old age range.

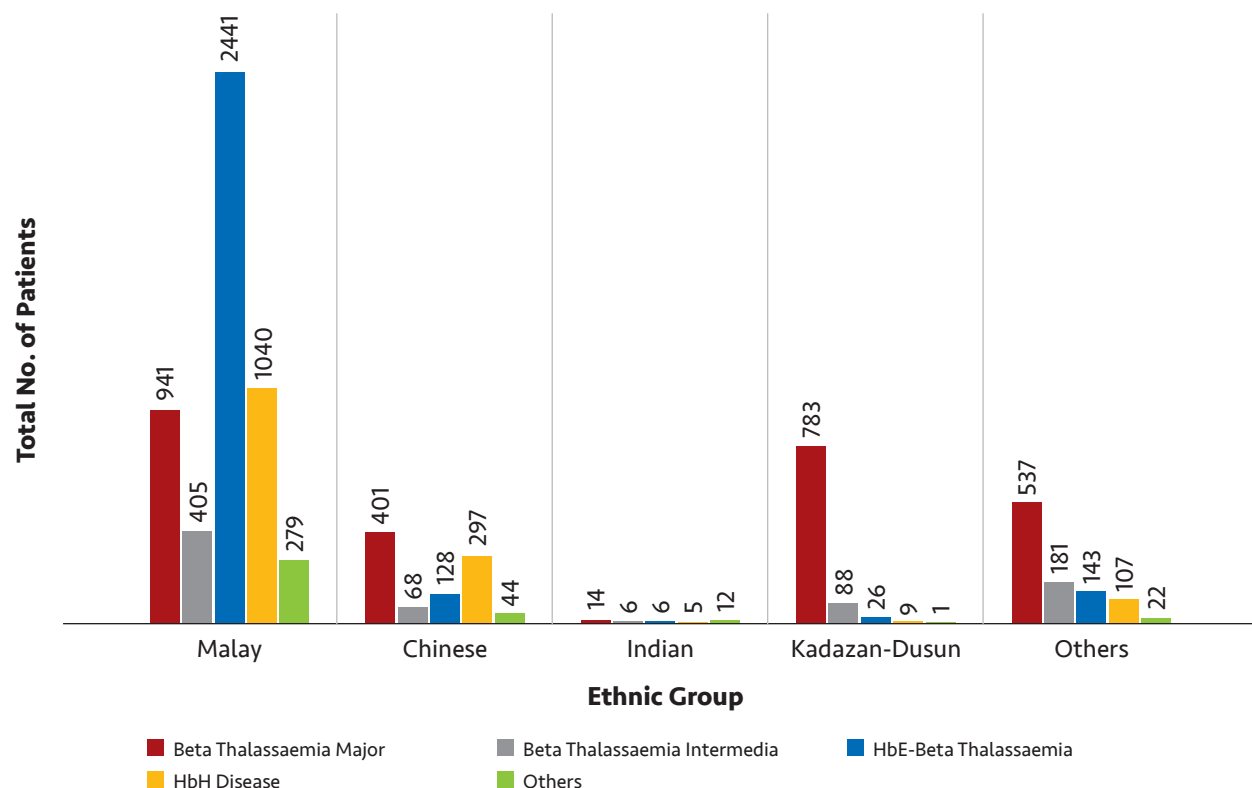
Table 2.17: Distribution of Patients According to Ethnic Group by Diagnosis

Diagnosis	Malay		Chinese		Indian		Kadazan-Dusun		Others	
	No.	%*	No.	%*	No.	%*	No.	%*	No.	%*
Beta Thalassaemia Major	941	18.43	401	42.75	14	32.56	783	86.33	537	54.24
Beta Thalassaemia Intermedia	405	7.93	68	7.25	6	13.95	88	9.70	181	18.28
HbE-Beta Thalassaemia	2441	47.81	128	13.65	6	13.95	26	2.87	143	14.44
HbH Disease	1040	20.37	297	31.66	5	11.63	9	0.99	107	10.81
Others	279	5.46	44	4.69	12	27.91	1	0.11	22	2.22
Total	5106	100.00	938	100.00	43	100.00	907	100.00	990	100.00

*Percentage are proportional to each total ethnic group respectively.

Table 2.17 demonstrates that HbE-beta thalassaemia was found to be more common among the Malay patients with 2441 patients (47.81%). Amongst the Chinese, Indian and Kadazan-Dusun, beta thalassaemia major was found to be most common within the ethnicities, with 401 (42.75%), 14 (32.56%), and 783 (86.33%) patients, respectively. Although thalassaemia is the most common genetic disorder in Malaysia, there is still scanty information describing thalassaemia in the other indigenous ethnic population.

Figure 2.16: Percentage Breakdown of Diagnoses by Ethnic Group



2.7 Iron Chelation Therapy

The management of severe forms of thalassaemia entails regular blood transfusion with chelation therapy to prevent the effects of iron accumulation. Iron removal following transfusional iron overload involves chelating drugs such as desferrioxamine (DFO), deferriprone (DFP) and deferasirox (DFX), or a combination of two or even all three of these chelating agents. Effective chelation therapy in chronically-transfused patients is achieved when iron chelators remove sufficient amounts of iron, equivalent to that accumulated in the body from transfusion, and maintain the body iron load at a non-toxic level.

As a general rule, transfusion-dependent thalassaemia (TDT) patients should start iron chelation therapy once they received 10-20 transfusions or when ferritin level rises above 1000 ng/mL. Iron chelating drugs have to be administered daily in an adequate dose.

DFO is a hexadentate iron chelator that binds iron in a 1:1 complex. DFO cannot be orally absorbed; therefore, it is administered subcutaneously or intravenously at a dose of 20–50 mg/kg/day, using a portable infusion pump. Its dosage for children is usually below 40 mg/kg. Higher doses of up to 60 mg/kg/day have been used in patients with high body iron stores. DFP was the first oral iron chelator to be used. It is approved in Europe and other countries for transfusional iron overload in patients with TDT, when DFO therapy is contraindicated or inadequate. It has a short half-life and needs to be taken three times daily. DFX, a new oral chelator, is a tridentate iron chelator that forms a 2:1 complex, with a once-daily dosing at 20–40 mg/kg/day. The plasma half-life of DFX is 16–18 hours, and it is predominantly excreted in biliary secretions.

Combination therapy with DFO and DFP was introduced to manage iron overload in patients who are sub-optimally chelated with maximum doses of DFP. The synergistic effect of DFP and DFO on iron balance and urine iron excretion has been explained by the shuttle mechanism. Combined chelation with DFP and DFX has also shown promising results and has been used for intensive chelation therapy. Triple combination therapy is seldom prescribed and it is advisable to check on patient adherence first since that is the most common cause of ineffective chelation.

From a total of 7984 patients, 4928 patients (61.72%) are receiving iron chelation therapy. Based on Table 2.18, 963 (19.54%), 1107 (22.46%) and 1645 (33.38%) patients are on monotherapy with DFO, DFP and DFX, respectively. DFX is normally reserved for chelator-naïve patients and for those who could not tolerate other chelating agents. The most common combination therapy was DFO/DFP with 910 patients (18.47%). Other combinations used were DFP/DFX and DFO/DFX, with 92 (1.87%) and 169 (3.43%) patients, respectively. There are 42 patients (0.85%) who are on DFO/DFP/DFX combination therapy.

Table 2.18: Distribution of Patients According to Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	963	19.54
DFP only	1107	22.46
DFX only	1645	33.38
DFO + DFP	910	18.47
DFP + DFX	92	1.87
DFO + DFX	169	3.43
DFO + DFP + DFX	42	0.85
Total	4928	100.00

Figure 2.17: Distribution of Patients According to Type of Iron Chelator Received

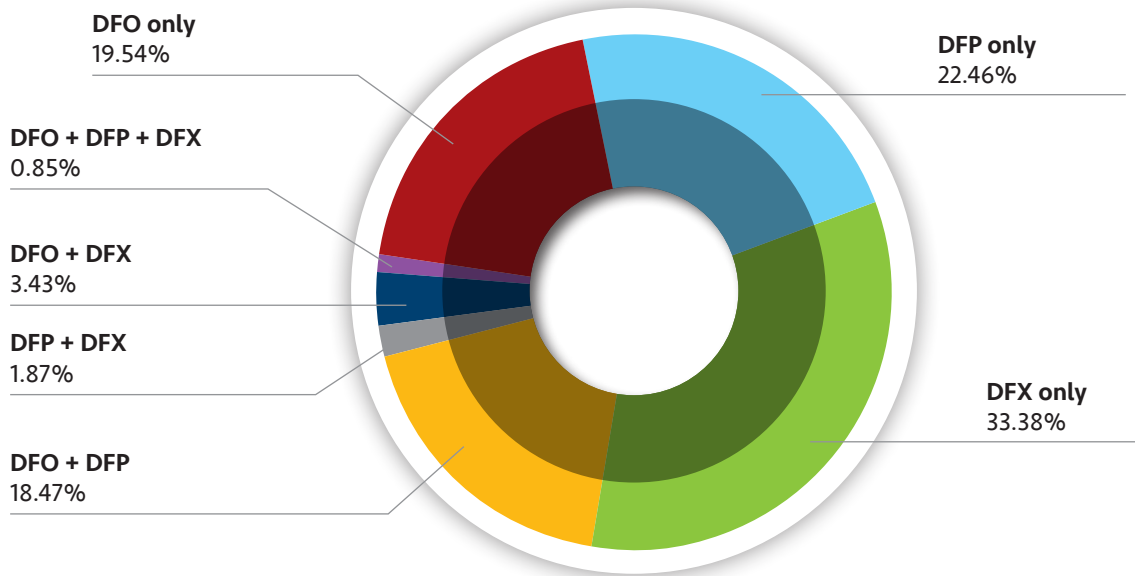
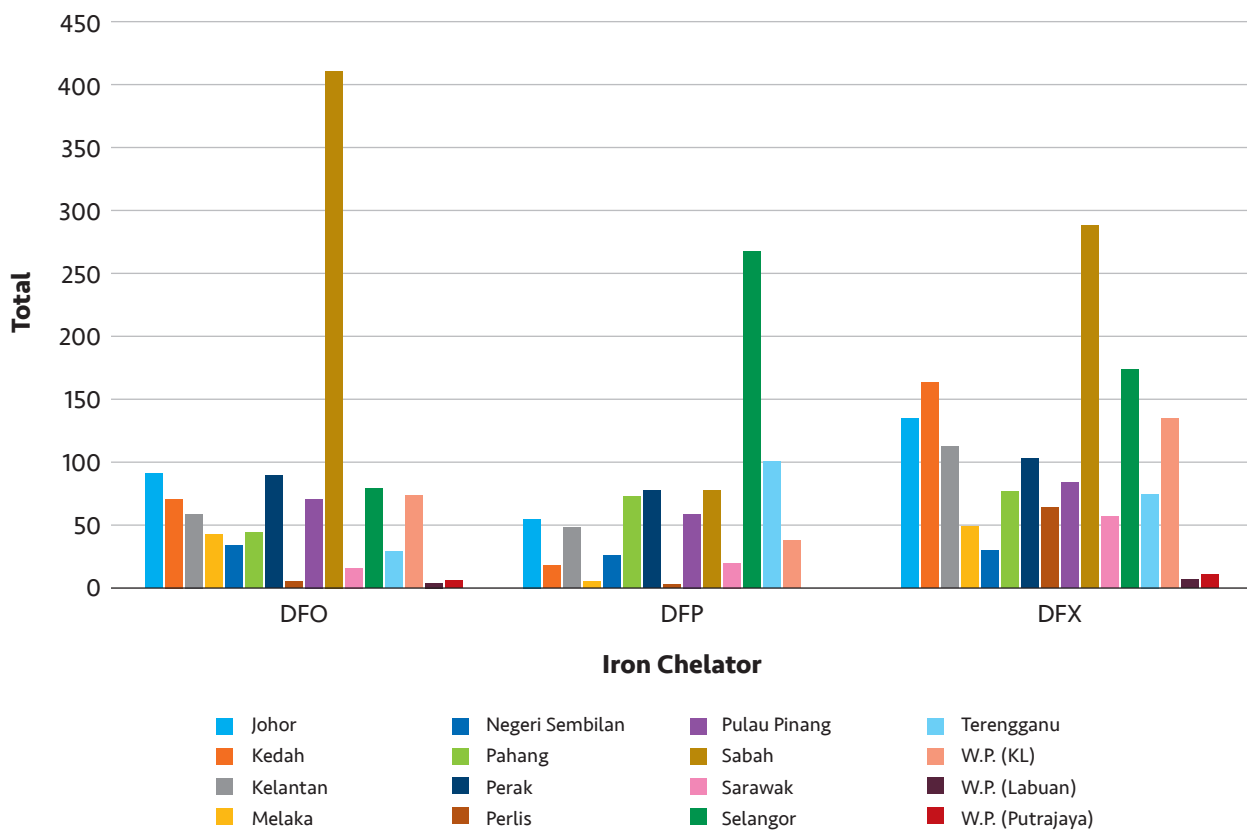


Figure 2.18: Distribution of Patients According to Type of Iron Chelator Received by State in 2017



2 Registry Report

Table 2.19: Distribution of Patients According to Type of Iron Chelator Received by State

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Johor	422	DFO only	63	14.93
		DFP only	119	28.20
		DFX only	112	26.54
		DFO + DFP	101	23.93
		DFP + DFX	8	1.90
		DFO + DFX	17	4.03
		DFO + DFP + DFX	2	0.47
Kedah	371	DFO only	36	9.70
		DFP only	70	18.87
		DFX only	200	53.91
		DFO + DFP	42	11.32
		DFP + DFX	11	2.96
		DFO + DFX	11	2.96
		DFO + DFP + DFX	1	0.27
Kelantan	276	DFO only	49	17.75
		DFP only	54	19.57
		DFX only	134	48.55
		DFO + DFP	18	6.52
		DFP + DFX	6	2.17
		DFO + DFX	14	5.07
		DFO + DFP + DFX	1	0.36
Melaka	115	DFO only	44	38.26
		DFP only	5	4.35
		DFX only	50	43.48
		DFO + DFP	12	10.43
		DFP + DFX	0	0.00
		DFO + DFX	4	3.48
		DFO + DFP + DFX	0	0.00
Negeri Sembilan	115	DFO only	33	28.70
		DFP only	27	23.48
		DFX only	31	26.96
		DFO + DFP	20	17.39
		DFP + DFX	0	0.00
		DFO + DFX	4	3.48
		DFO + DFP + DFX	0	0.00

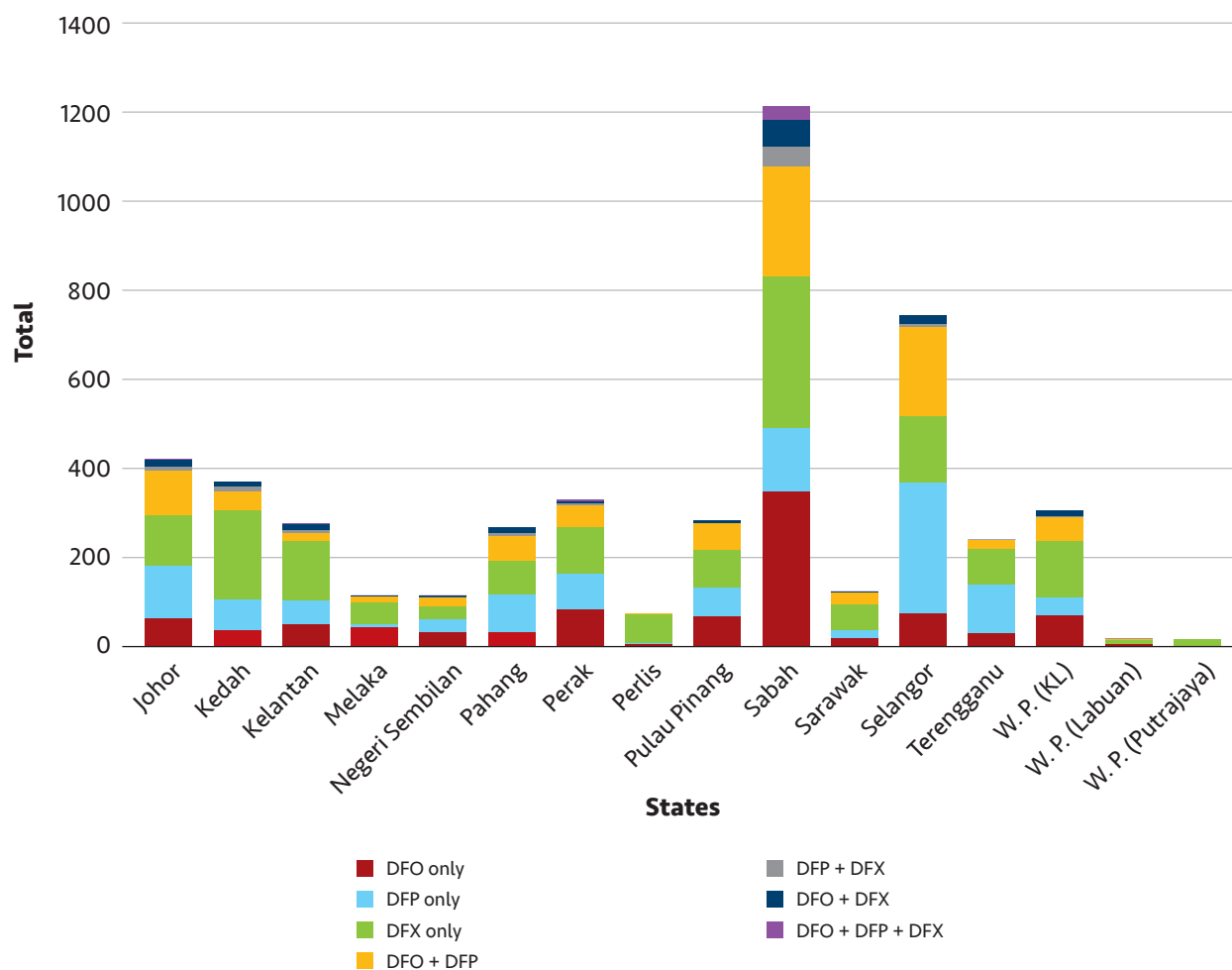
Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Pahang	267	DFO only	33	12.36
		DFP only	83	31.09
		DFX only	77	28.84
		DFO + DFP	55	20.60
		DFP + DFX	5	1.87
		DFO + DFX	14	5.24
		DFO + DFP + DFX	0	0.00
Perak	329	DFO only	84	25.53
		DFP only	78	23.71
		DFX only	105	31.91
		DFO + DFP	49	14.89
		DFP + DFX	6	1.82
		DFO + DFX	4	1.22
		DFO + DFP + DFX	3	0.91
Perlis	74	DFO only	5	6.76
		DFP only	2	2.70
		DFX only	65	87.84
		DFO + DFP	2	2.70
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Pulau Pinang	283	DFO only	68	24.03
		DFP only	64	22.61
		DFX only	85	30.04
		DFO + DFP	60	21.20
		DFP + DFX	0	0.00
		DFO + DFX	6	2.12
		DFO + DFP + DFX	0	0.00
Sabah	1214	DFO only	349	28.75
		DFP only	142	11.70
		DFX only	339	27.92
		DFO + DFP	248	20.43
		DFP + DFX	44	3.62
		DFO + DFX	59	4.86
		DFO + DFP + DFX	33	2.72

2 Registry Report

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Sarawak	124	DFO only	18	14.52
		DFP only	19	15.32
		DFX only	56	45.16
		DFO + DFP	27	21.77
		DFP + DFX	0	0.00
		DFO + DFX	4	3.23
		DFO + DFP + DFX	0	0.00
Selangor	753	DFO only	75	9.58
		DFP only	292	37.29
		DFX only	161	20.56
		DFO + DFP	198	25.29
		DFP + DFX	8	1.02
		DFO + DFX	18	2.30
		DFO + DFP + DFX	1	0.13
Terengganu	240	DFO only	29	12.08
		DFP only	111	46.25
		DFX only	78	32.50
		DFO + DFP	22	9.17
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
WP Kuala Lumpur	306	DFO only	70	21.88
		DFP only	39	12.19
		DFX only	128	40.00
		DFO + DFP	53	16.56
		DFP + DFX	2	0.63
		DFO + DFX	14	4.38
		DFO + DFP + DFX	0	0.00
WP Labuan	18	DFO only	3	15.79
		DFP only	0	0.00
		DFX only	9	47.37
		DFO + DFP	3	15.79
		DFP + DFX	2	10.53
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	5.26

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
WP Putrajaya	21	DFO only	4	18.18
		DFP only	2	9.09
		DFX only	15	68.18
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			4928	100.00

Figure 2.19: Distribution of Patients According to Type of Iron Chelator Received by State



2 Registry Report

Table 2.20: Distribution of Patients According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	187	DFO only	12	0.24
		DFP only	4	0.08
		DFX only	166	3.37
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	5	0.10
		DFO + DFP + DFX	0	0.00
5-9.9	739	DFO only	70	1.42
		DFP only	39	0.79
		DFX only	559	11.34
		DFO + DFP	13	0.26
		DFP + DFX	22	0.45
		DFO + DFX	33	0.67
		DFO + DFP + DFX	3	0.06
10-14.9	910	DFO only	133	2.70
		DFP only	127	2.58
		DFX only	506	10.27
		DFO + DFP	75	1.52
		DFP + DFX	22	0.45
		DFO + DFX	40	0.81
		DFO + DFP + DFX	7	0.14
15-19.9	874	DFO only	257	5.22
		DFP only	165	3.35
		DFX only	224	4.55
		DFO + DFP	176	3.57
		DFP + DFX	17	0.34
		DFO + DFX	31	0.63
		DFO + DFP + DFX	4	0.08
20-24.9	818	DFO only	237	4.81
		DFP only	157	3.19
		DFX only	94	1.91
		DFO + DFP	274	5.56
		DFP + DFX	12	0.24
		DFO + DFX	31	0.63
		DFO + DFP + DFX	13	0.26

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
25-29.9	512	DFO only	121	2.46
		DFP only	158	3.21
		DFX only	36	0.73
		DFO + DFP	163	3.31
		DFP + DFX	9	0.18
		DFO + DFX	15	0.30
		DFO + DFP + DFX	10	0.20
30-34.9	351	DFO only	66	1.34
		DFP only	145	2.94
		DFX only	23	0.47
		DFO + DFP	102	2.07
		DFP + DFX	4	0.08
		DFO + DFX	7	0.14
		DFO + DFP + DFX	4	0.08
35-39.9	186	DFO only	25	0.51
		DFP only	97	1.97
		DFX only	9	0.18
		DFO + DFP	50	1.01
		DFP + DFX	2	0.04
		DFO + DFX	2	0.04
		DFO + DFP + DFX	1	0.02
40-44.9	122	DFO only	12	0.24
		DFP only	80	1.62
		DFX only	6	0.12
		DFO + DFP	21	0.43
		DFP + DFX	1	0.02
		DFO + DFX	2	0.04
		DFO + DFP + DFX	0	0.00
45-49.9	94	DFO only	15	0.30
		DFP only	53	1.08
		DFX only	9	0.18
		DFO + DFP	14	0.28
		DFP + DFX	1	0.02
		DFO + DFX	2	0.04
		DFO + DFP + DFX	0	0.00

2 Registry Report

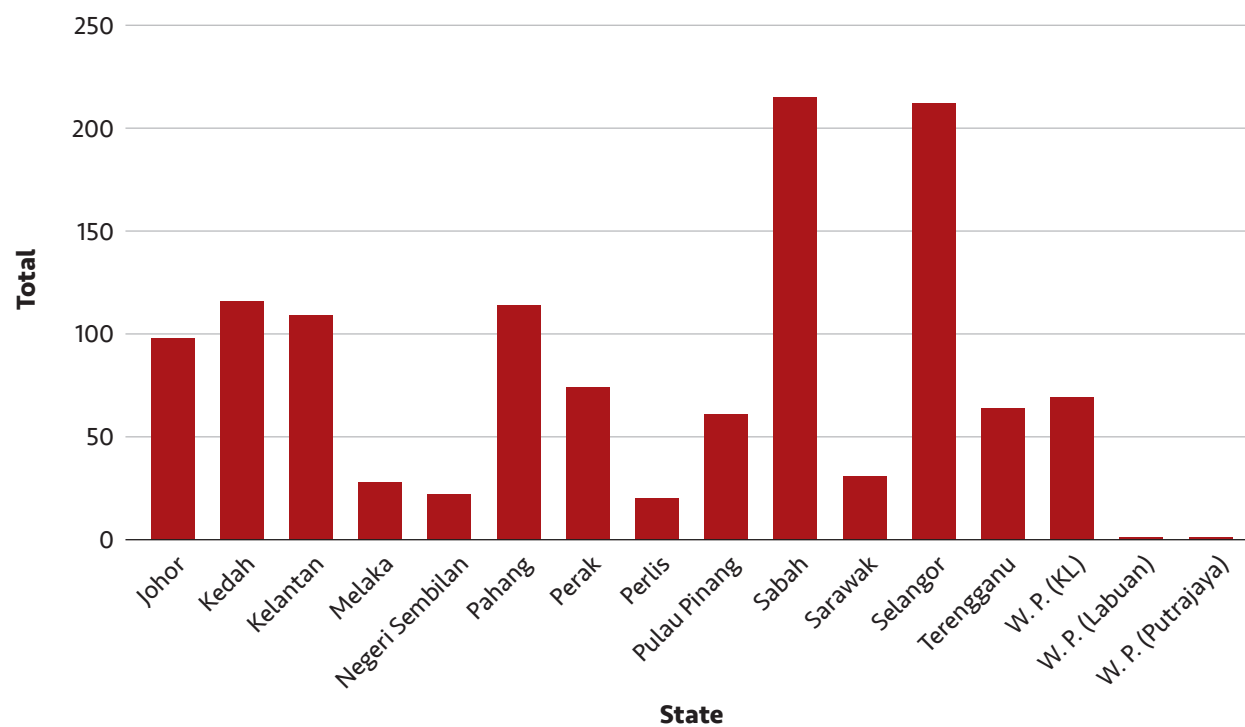
Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
50-54.9	53	DFO only	7	0.14
		DFP only	32	0.65
		DFX only	2	0.04
		DFO + DFP	11	0.22
		DFP + DFX	1	0.02
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	31	DFO only	4	0.08
		DFP only	16	0.32
		DFX only	2	0.04
		DFO + DFP	7	0.14
		DFP + DFX	1	0.02
		DFO + DFX	1	0.02
		DFO + DFP + DFX	0	0.00
60-64.9	27	DFO only	1	0.02
		DFP only	20	0.41
		DFX only	4	0.08
		DFO + DFP	2	0.04
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
> 65	24	DFO only	3	0.06
		DFP only	14	0.28
		DFX only	5	0.10
		DFO + DFP	2	0.04
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			4928	100.00

2.8 Splenectomy

The major reason for undergoing splenectomy is hypersplenism. For patients with thalassaemia in whom yearly transfusion requirements exceed 200 mL packed cells per kilogram body weight, a splenectomy should significantly diminish red blood cell requirements and iron accumulation (Modell, 1977; Cohen, 1989). Hypersplenism may be avoided by early and regular transfusion; many patients reaching adolescence in this decade have not required splenectomy. Because of the risk of post-splenectomy infection, splenectomy should generally be delayed until the age of 5 or later. From a total of 8681 patients in Malaysia, only 1235 patients (14.23%) have undergone splenectomy.

Table 2.21: Distribution of Splenectomised Patients by State

State	Total No. of Patients	No. of Splenectomised Patients	Percentage (%)
Johor	667	98	1.13
Kedah	725	116	1.34
Kelantan	515	109	1.26
Melaka	228	28	0.32
Negeri Sembilan	187	22	0.25
Pahang	473	114	1.31
Perak	608	74	0.85
Perlis	140	20	0.23
Pulau Pinang	492	61	0.70
Sabah	2117	215	2.48
Sarawak	244	31	0.36
Selangor	1233	212	2.44
Terengganu	367	64	0.74
WP (KL)	614	69	0.79
WP (Labuan)	29	1	0.01
WP (Putrajaya)	42	1	0.01
Total	8681	1235	14.23

Figure 2.20: Distribution of Splenectomised Patients by State

2 Registry Report

2.9 Regular Transfusion

Regular blood transfusion will mitigate the effects of chronic anaemia in TDT and other refractory anaemias. However, transfusional iron overload would lead to chronic iron toxicities, multiple organ dysfunction and eventually death. Patients who are left untreated, including by both transfusions and chelating drugs, have poor survival rates and high morbidity and mortality, as reported in other countries. The amount of blood to be transfused depends on several factors including the patient's weight and the target increase in Hb level. Ideally the Hb levels should always be kept above 9 g/dL.

In Malaysia, there are 4529 TDT patients who receive regular packed cell transfusion 3-4 times weekly to alleviate the chronic anaemia that is associated with the disease. Regular transfusion is defined as receiving a minimum of packed cells transfusion at least on a 6-weekly interval.

Table 2.22: Distribution of Patients Receiving Regular Transfusions by State

State	Total No. of Patients	No. of Patients Receiving Regular Transfusion	Percentage (%)	No. of Transfusions
Johor	637	395	8.72	2643
Kedah	694	341	7.53	1209
Kelantan	486	271	5.98	1842
Melaka	226	105	2.32	531
Negeri Sembilan	181	101	2.23	488
Pahang	437	222	4.90	1084
Perak	564	246	5.43	837
Perlis	128	75	1.66	112
Pulau Pinang	480	226	4.99	928
Sabah	1814	1216	26.85	4805
Sarawak	223	111	2.45	1026
Selangor	1169	600	13.25	3300
Terengganu	344	245	5.41	1937
WP (Kuala Lumpur)	535	331	7.31	1242
WP (Labuan)	25	18	0.40	78
WP (Putrajaya)	41	26	0.57	171
Total	7984	4529	100.00	22233

Figure 2.21: Distribution of Patients Receiving Regular Transfusions by State

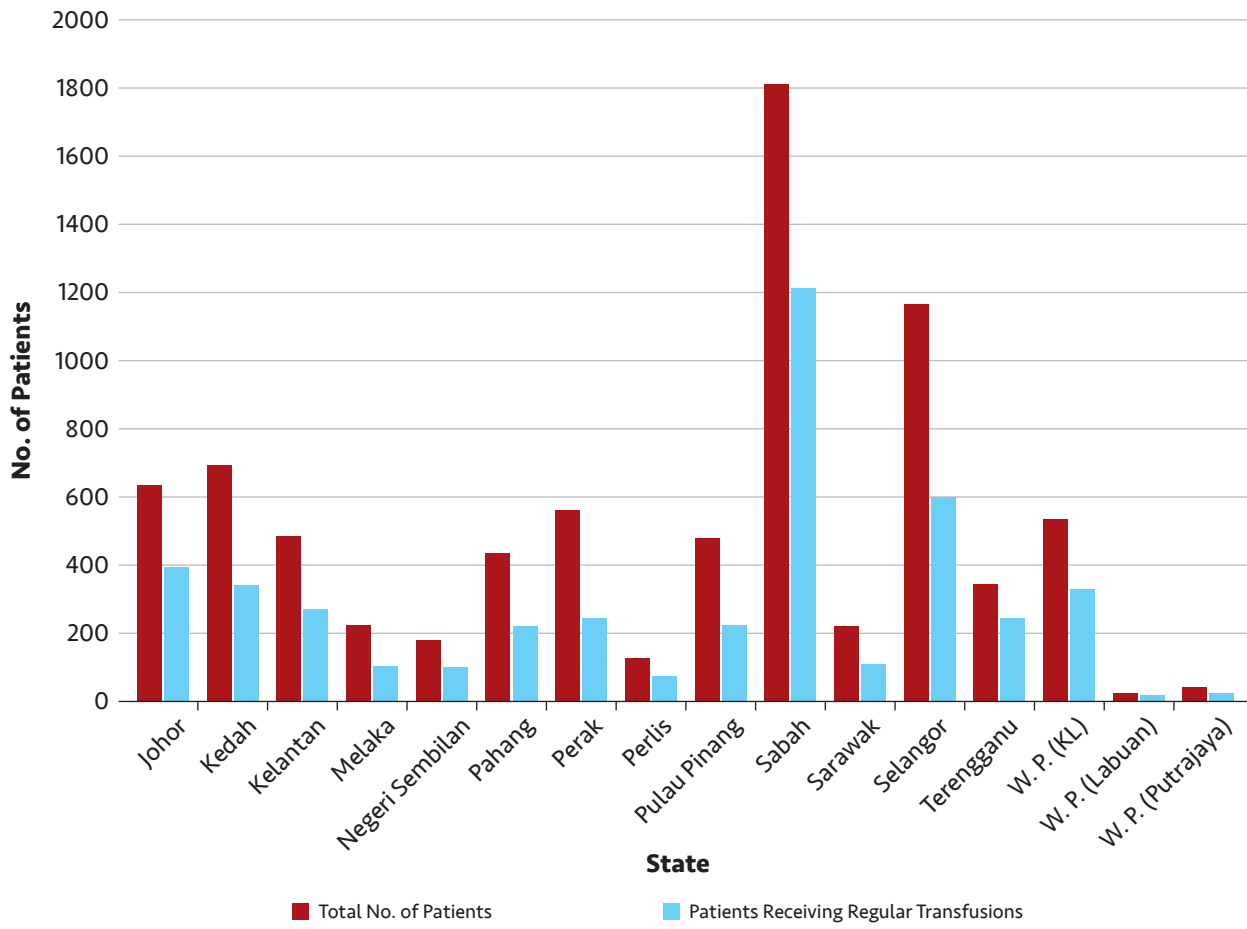
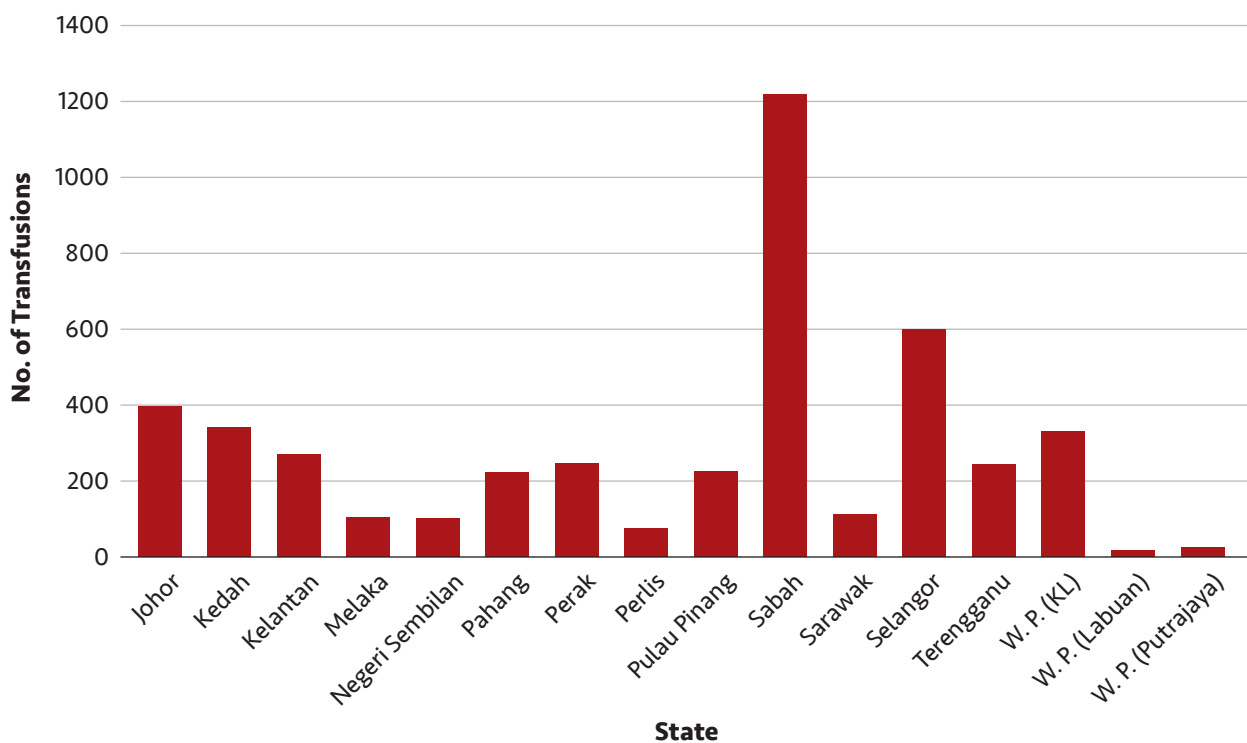


Figure 2.22: Total Number of Transfusions by State



2 Registry Report

Table 2.23: Total Number of Patients Receiving Regular Transfusions by Year

Year	Total No. of Patients	No. of Patients Receiving Regular Transfusion	No. of Transfusions
2018	7984	4529	22233
2017	7882	4490	25708
2016	7605	4370	20621
2015	7217	4203	17292
2014	6805	4018	15201
2013	6386	3832	14796
2012	5973	3628	14843
2011	5547	3401	12911
2010	5164	3202	16885

56

Based on Table 2.23, there is under-reporting of transfusion received in the registry. However, the number of blood transfusions can be seen to be increasing annually. This is due to the increasing number of patients as well as their weights as they grow older. In addition, liver and spleen sizes also indirectly indicate the adequacy of blood transfusion (Mutha *et al.*, 2018). Thus, an increase in liver and spleen sizes might indicate extramedullary haematopoiesis and more blood transfusion is required to control the size of these organs. However, there are risks of regular transfusion including multi-organ haemosiderosis and blood-borne infections (Chan *et al.*, 2001). Therefore, careful monitoring of blood transfusion management and regular screening of transfusion-related infections need to be undertaken.

2.10 Serum Ferritin Level

Iron overload is the most problematic complication associated with chronic transfusion therapy. Patients on regular transfusion regimen will progressively develop clinical manifestations unless treated promptly and adequately. Thus, iron overload status should be accurately and continuously assessed to evaluate its severity, the need for treatment, timing and monitoring of chelation therapy. Serum ferritin level has in general been found to correlate well with body iron store. Although high ferritin level represents iron overload, this positive acute phase protein may also be increased during acute and chronic disorders. Thus, two more sensitive and specific parameters of tissue iron overload, which are T2* liver and cardiac magnetic resonance imaging (MRI) should also be performed (Chirico *et al.*, 2015).

There is a decreasing number of patients who have a high serum ferritin level beyond 5,000 ng/mL. Correspondingly, more patients are now having serum ferritin levels below 2500 ng/mL. This would indicate an improvement in iron chelating management and should translate into better long-term outcome and preservation of vital organ functions.

Table 2.24: Distribution of Patients According to Most Recent Serum Ferritin Levels by State in 2018

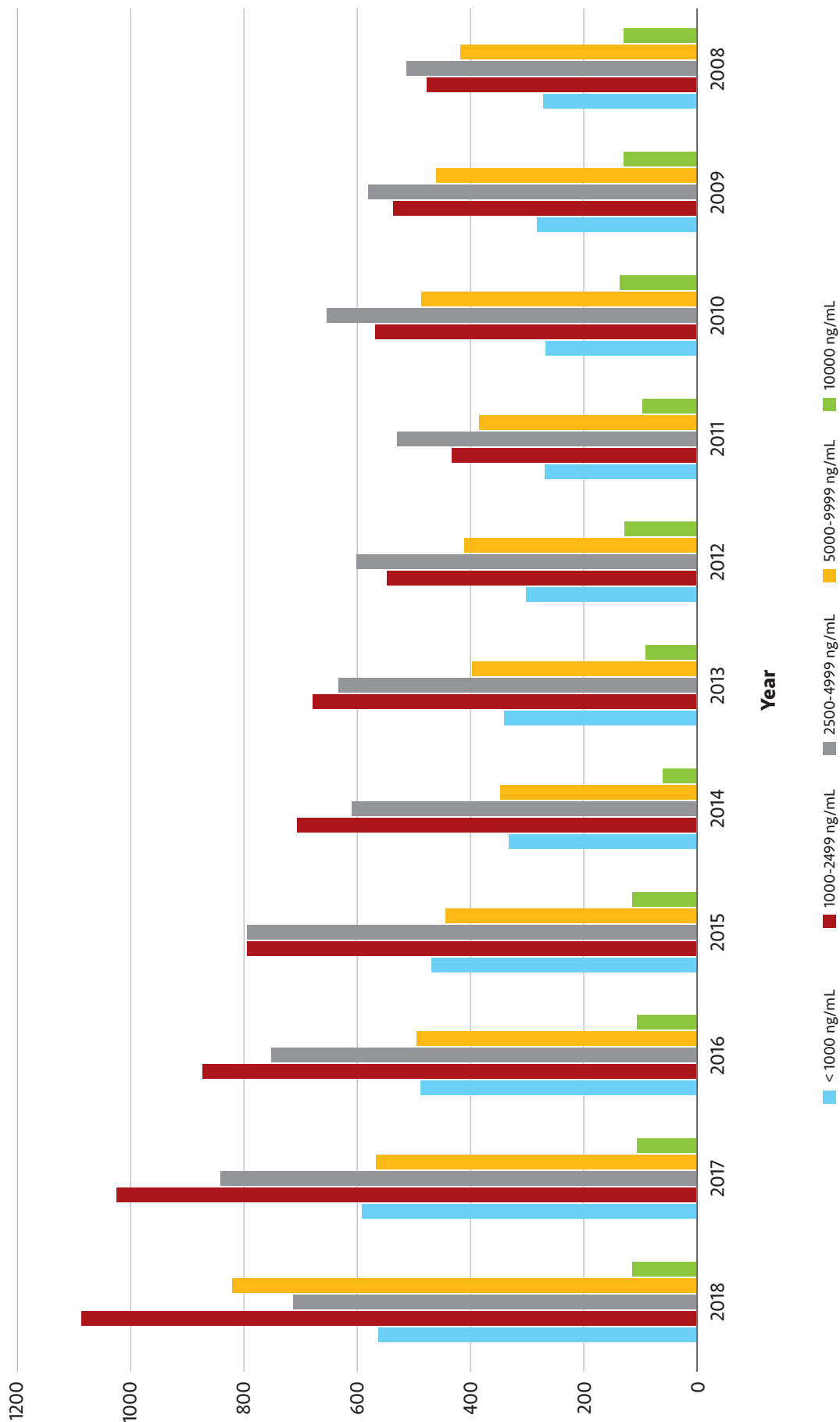
Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
State	Total No. of Patients	No.	%*	No.	%*	No.	%*	No.	%*	No.	%*
Johor	326	53	16.26	90	27.61	108	33.13	64	19.63	11	3.37
Kedah	266	48	18.05	102	38.35	71	26.69	33	12.41	12	4.51
Kelantan	210	43	20.48	81	38.57	52	24.76	26	12.38	8	3.81
Melaka	42	6	14.29	20	47.62	10	23.81	5	11.90	1	2.38
N. Sembilan	61	8	13.11	16	26.23	23	37.70	8	13.11	6	9.84
Pahang	152	29	19.08	69	45.39	27	17.76	23	15.13	4	2.63
Perak	181	34	18.78	59	32.60	52	28.73	28	15.47	8	4.42
Perlis	33	5	15.15	13	39.39	10	30.30	5	15.15	0	0.00
P. Pinang	127	24	18.90	42	33.07	36	28.35	23	18.11	2	1.57
Sabah	588	88	14.97	181	30.78	150	25.51	148	25.17	21	3.57
Sarawak	100	29	29.00	39	39.00	19	19.00	10	10.00	3	3.00
Selangor	480	62	12.92	183	38.13	131	27.29	80	16.67	24	5.00
Terengganu	221	61	27.60	74	33.48	54	24.43	27	12.22	5	2.26
KL	267	63	23.60	101	37.83	68	25.47	30	11.24	5	1.87
Labuan	13	3	23.08	3	23.08	6	46.15	0	0.00	1	7.69
Putrajaya	24	6	25.00	14	58.33	3	12.50	1	4.17	0	0.00
Total	3091	562	18.18	1087	35.17	820	23.03	511	16.53	111	3.59

*Percentage is calculated against number of patients for the respective state.

Table 2.25: Distribution of Patients According to Serum Ferritin Levels by Year

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
State	Total No. of Patients	No.	%	No.	%	No.	%	No.	%	No.	%
2018	3091	562	18.18	1087	35.17	820	26.53	511	16.53	111	3.59
2017	3126	591	18.91	1024	32.76	841	26.90	566	18.11	104	3.33
2016	2709	487	17.98	872	32.19	751	27.72	494	18.24	105	3.88
2015	2612	468	17.92	794	30.40	794	30.40	443	16.96	113	4.33
2014	2051	331	16.14	706	34.42	609	29.69	346	16.87	59	2.88
2013	2134	339	15.89	678	31.77	632	29.62	396	18.56	89	4.17
2012	1984	301	15.17	546	27.52	601	30.29	410	20.67	126	6.35
2011	1706	268	15.71	432	25.32	528	30.95	384	22.51	94	5.51
2010	2107	266	12.62	567	26.91	653	30.99	486	23.07	135	6.41
2009	1982	281	14.18	535	26.99	579	29.21	459	23.16	128	6.46
2008	1803	270	14.98	476	26.40	512	28.40	417	23.13	128	7.10
2007	1522	261	17.15	406	26.68	391	25.69	339	22.27	125	8.21

Figure 2.23: Distribution of Patients According to Serum Ferritin Levels by Year



3 Conclusions and Observations

Prior to the development of the MTR, the prevalence of the thalassaemia syndromes in Malaysia were based on MOH annual reports, news archives, small studies, single-centred population or reports focused mainly on molecular characterisation or evaluation of quality of life according to state, centre, or ethnicity. Although the significance of thalassaemia in the region was emphasised, these sources lacked the exact figure of reported cases in the respective regions where the study was conducted.

As demonstrated in this report, the total number of thalassaemia patients in the registry has significantly rose over the years, totalling 7984 patients in 2018. The increasing numbers were due to improved survival, new births of thalassaemia patients as well as more newly diagnosed patients with less severe forms of thalassaemia being registered in the MTR. The initial estimated number of patients were 2500 nationwide in 2004, according to a survey sent out to major government hospitals treating TDT patients. With the launch of a web-based registry, more centres can now submit data online and currently a total of 110 centres are participating.

Although there appears to be a decreasing trend in the number of new thalassaemia births registered, the total numbers are still significant and more needs to be done to reduce it even further. Special emphasis should be given to Sabah as it has the highest numbers of registered patients and new thalassaemia births. Preventive measures, such as the targeted screening of all 16-year-olds since November 2016 should be followed up with screening and counselling of young and pre-marital couples. Provision of prenatal diagnostic services and selective termination of affected fetuses would also need to be developed further.

The most common diagnosis was HbE-beta thalassaemia with 2744 patients, followed by beta thalassaemia major (2676 patients) and HbH disease (1458 patients). Malays predominantly have a higher prevalence of HbE-beta thalassaemia, with the highest proportion found in Selangor, Kuala Lumpur, Kelantan, and Kedah. Beta thalassaemia major is especially predominant among the Kadazan-Dusuns of Sabah due to the high prevalence of Filipino beta thalassaemia deletion in the ethnicity. Incidentally, HbH disease, which is commonly caused by the deletion of three alpha-globin genes ($-\alpha/\alpha$), are more common among the Malaysian Chinese.

Clinicians are encouraged to prescribe monotherapy whenever possible unless the chelation is inadequate, or patients are not compliant. An example is poorer compliance or tolerance towards DFO, which is administered subcutaneously for 8-12 hours, leading to the change to oral chelators such as DFX or DFP. DFX is the more popular option as it can be prescribed to younger patients (minimum 2 years old, in the form of dispersible tablets, once daily) compared to DFP (6 years and older, in the form of tablets or capsule, thrice daily). Combination therapy, in contrast, will only be initiated when intensive therapy is needed to overcome severe iron overload.

Iron chelation in the current population has shown a positive impact in decreasing the serum ferritin level. Although high-risk patients with levels above 10000 ng/mL are still present, the decline in their numbers indicate improvement in patient management. There is also appreciable decline in those with levels beyond 5000 ng/mL as well as an increase in those with levels below 2500 ng/mL. Nevertheless, cardiac complications due to iron overload is still the leading cause of death among thalassaemia patients. Thus, participating centres will need to improve on iron overload monitoring and chelation therapy, especially in high-risk patients. In addition, assessment using ferritin level alone is not specific and would not identify those with tissue iron deposition especially in the heart and liver. Therefore, there is an urgent need to expand further the routine use of magnetic resonance imaging (MRI) T2* to assess iron deposition in tissues. Currently, the MRI T2* data collection for the MTR has yet to begin and we hope to address this in future.

Although the MTR was able to gather most information on thalassaemia patients nationwide, certain records are still incomplete. Among the notable limitations are serum ferritin levels, death records, and birth summary. Certain information might not be accessible, or the evaluation was not completed on the whole population, leading to missing information. Data on thalassaemia births in 2017 and 2018, for instance, may not be up-to-date since disease presentation appears later during infancy or even later than that for patients with thalassaemia intermedia, and will be registered only after disease manifestation. Laboratory data on serum ferritin, on the other hand, might not be recorded in the patient's notes and thus unavailable for analysis.

This MTR report describes the current and evolving scenario of thalassaemia in Malaysia over the past decade. The landscape has changed dramatically as patients are now living past the second decade of life with improvement of patient management. Data gathered into the MTR can be used to understand the progression of the disorder and to improve treatment outcomes, develop strategies on iron overload management, formulate policies and preventive strategies to reduce the healthcare burden of thalassaemia.

4 Johor

4.1 Introduction

Johor is located in the southern part of Peninsular Malaysia. Based on the latest data from the Department of Statistics Malaysia for the year 2018, the total population of the state of Johor is about 3.742 million people.

There are 12 hospitals in the state of Johor. Six are hospitals with specialists, namely Hospital Sultanah Aminah (Johor Bahru), Hospital Sultan Ismail (Johor Bahru), Hospital Enche' Besar Hajjah Khalsom (Kluang), Hospital Sultanah Nora Ismail (Batu Pahat), Hospital Pakar Sultanah Fatimah (Muar) and Hospital Segamat. On the other hand, another five hospitals, namely Hospital Temenggong Seri Maharaja Tun Ibrahim (Kulai), Hospital Kota Tinggi, Hospital Mersing, Hospital Pontian and Hospital Tangkak are district hospitals. Hospital Permai solely treats psychiatric patients.

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

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

4.2 Patient Demographics

There are currently 582 living patients in Johor, 14 of which were cured by stem cell transplantation. There were four deaths in 2018 and another 41 patients were lost to follow-up. A total of 78 new cases were reported in 2018. As of today, there is no new thalassaemia birth reported in 2018; however, there were seven new births reported in 2017. Most of the new births are beta thalassaemia major. There were four deaths in 2018 due to severe anaemia, Pseudomonas septicaemia with history of splenectomy, dengue fever and hepatocellular carcinoma. Majority of patients received treatment at Hospital Sultanah Aminah (48.51%) and Hospital Sultan Ismail (14.60%), as both centres are located in Johor Bahru (which has the highest population in the state at about 1.3 million) and are the referral centres for thalassaemia cases.

Table 4.1: Distribution of Patients by Centre in Johor

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
Temenggong Seri Maharaja Tun Ibrahim, Kulaijaya	5	0.78
Sultanah Aminah, Johor Bahru	309	48.51
Pakar Sultanah Fatimah, Muar	74	11.62
Sultanah Nora Ismail, Batu Pahat	32	5.02
Enche' Besar Hajjah Kalsom, Kluang	47	7.38
Segamat	39	6.12
Pontian	7	1.10
Kota Tinggi	4	0.63
Mersing	27	4.24
Tangkak	0	0.00
Sultan Ismail, Johor Bahru	93	14.60
Total	637	100

4 Johor

Figure 4.1: Distribution of Patients by Centre in Johor

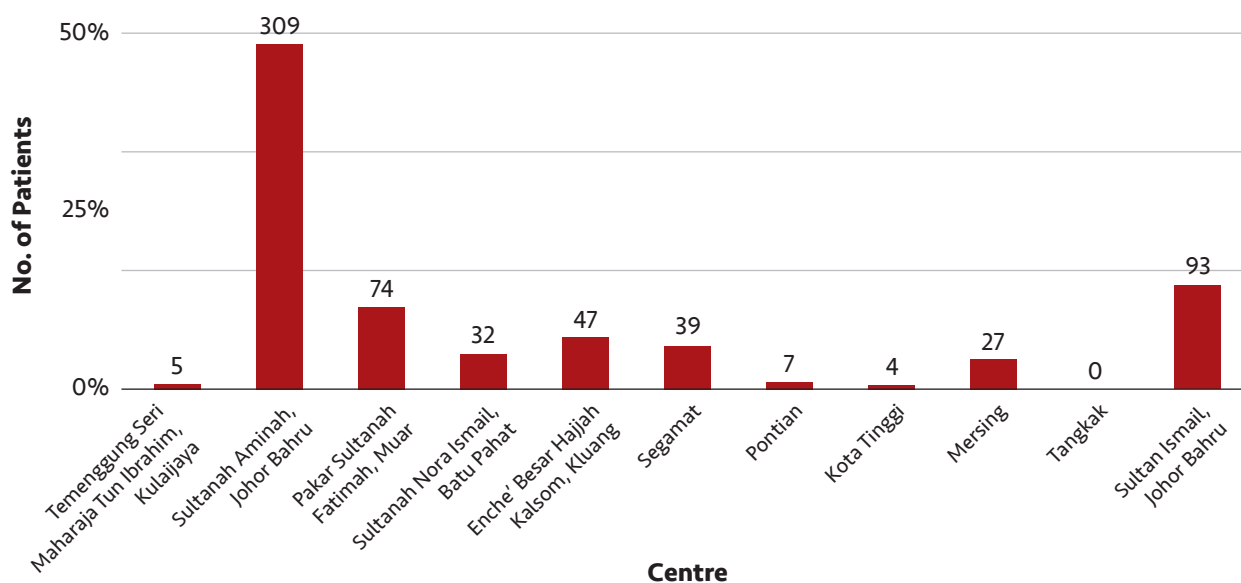


Table 4.2: Distribution of Patients by Vital Status in Johor

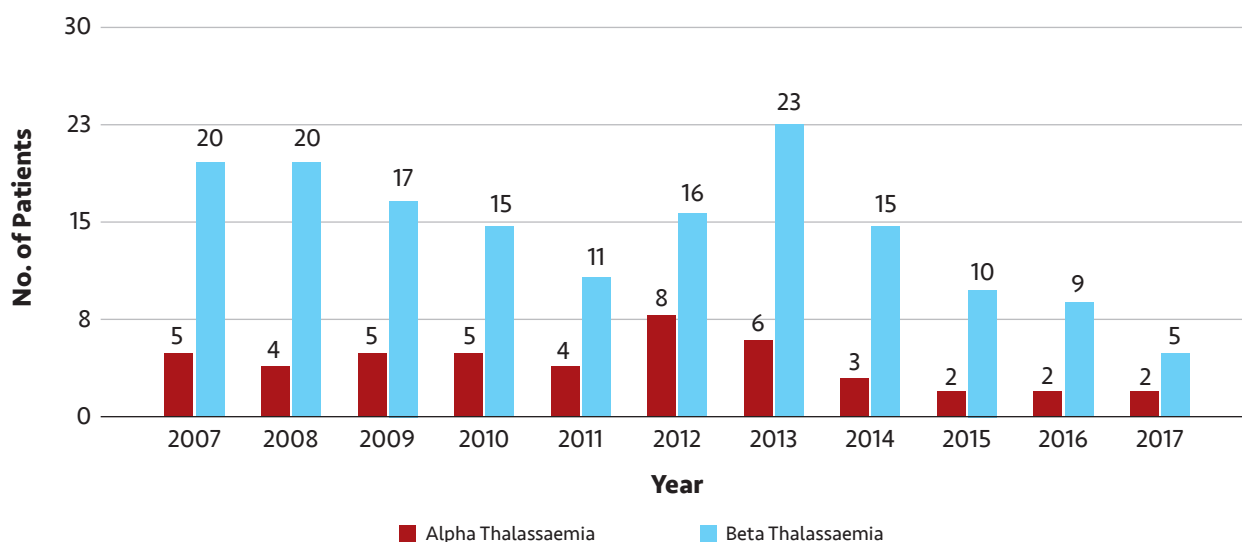
Vital Status	No. Of Patients
Alive	582
Cumulative Cured by Stem Cell Therapy	14
Lost to Follow-Up	41
Total	637
Deaths in 2018	4
Cumulative Reported Deaths (from 2009 to 2018)	30
Alive	582

Table 4.3: Cumulative Causes of Death Since 2007 in Johor

Cause of Death	No. of Patients
Infections	16
Cardiac & Anaemia	10
Multiple Organ Dysfunction	1
Tumour	2
Motor Vehicle Accident (MVA)	1
Unknown*	0
Total	30

*Missing data.

Figure 4.2: Distribution of New Thalassaemia Births in Johor, According to Type of Thalassaemia by Year



4.2.1 Age

The youngest patient recorded in Johor is a 1.5 year-old with HbE-beta thalassaemia (from Hospital Sultan Ismail) and the oldest is a 69 year-old with HbH disease. The oldest patient with beta thalassaemia major is 54 years old. Both the older patients are from Hospital Sultanah Aminah, Johor Bahru.

Table 4.4: Distribution of Patients in Johor by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	48	7.54
5 - 9.9	110	17.27
10 -14.9	101	15.86
15 -19.9	108	16.95
20 - 24.9	97	15.23
25 - 29.9	59	9.26
30 - 34.9	32	5.02
35 - 39.9	30	4.71
40 - 44.9	15	2.35
45 - 49.9	16	2.51
50 - 54.9	7	1.10
55 - 59.9	8	1.26
60 - 64.9	3	0.47
Above 65	3	0.47
Total	637	100.00

Figure 4.3: Distribution of Patients in Johor by Age Group

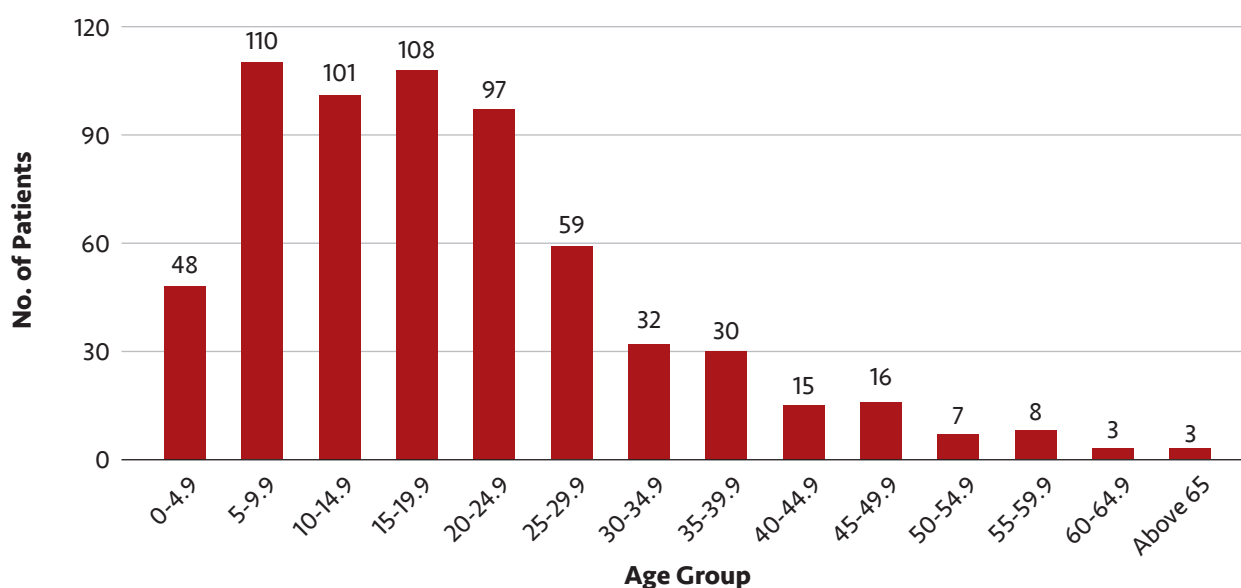


Table 4.5: Distribution of Patients in Johor According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	48	Beta Thalassaemia Major	16	2.51
		Beta Thalassaemia Intermedia	2	0.31
		HbE-Beta Thalassaemia	21	3.30
		HbH Disease	8	1.26
		Others*	1	0.16
5 - 9.9	110	Beta Thalassaemia Major	29	4.55
		Beta Thalassaemia Intermedia	7	1.10
		HbE-Beta Thalassaemia	46	7.22
		HbH Disease	27	4.24
		Others*	1	0.16
10 -14.9	101	Beta Thalassaemia Major	24	3.77
		Beta Thalassaemia Intermedia	8	1.26
		HbE-Beta Thalassaemia	51	8.01
		HbH Disease	18	2.83
		Others*	0	0.00
15 -19.9	108	Beta Thalassaemia Major	35	5.49
		Beta Thalassaemia Intermedia	4	0.63
		HbE-Beta Thalassaemia	47	7.38
		HbH Disease	21	3.30
		Others*	1	0.16

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
20 - 24.9	97	Beta Thalassaemia Major	41	6.44
		Beta Thalassaemia Intermedia	4	0.63
		HbE-Beta Thalassaemia	40	6.28
		HbH Disease	12	1.88
		Others*	0	0.00
25 - 29.9	59	Beta Thalassaemia Major	28	4.40
		Beta Thalassaemia Intermedia	2	0.31
		HbE-Beta Thalassaemia	25	3.92
		HbH Disease	4	0.63
		Others*	0	0.00
30 - 34.9	32	Beta Thalassaemia Major	15	2.35
		Beta Thalassaemia Intermedia	5	0.78
		HbE-Beta Thalassaemia	9	1.41
		HbH Disease	3	0.47
		Others*	0	0.00
35 - 39.9	30	Beta Thalassaemia Major	10	1.57
		Beta Thalassaemia Intermedia	9	1.41
		HbE-Beta Thalassaemia	6	0.94
		HbH Disease	5	0.78
		Others*	0	0.00
40 - 44.9	15	Beta Thalassaemia Major	2	0.31
		Beta Thalassaemia Intermedia	2	0.31
		HbE-Beta Thalassaemia	7	1.10
		HbH Disease	4	0.63
		Others*	0	0.00
45 - 49.9	16	Beta Thalassaemia Major	6	0.94
		Beta Thalassaemia Intermedia	4	0.63
		HbE-Beta Thalassaemia	5	0.78
		HbH Disease	1	0.16
		Others*	0	0.00
50 - 54.9	7	Beta Thalassaemia Major	2	0.31
		Beta Thalassaemia Intermedia	3	0.47
		HbE-Beta Thalassaemia	2	0.31
		HbH Disease	0	0.00
		Others*	0	0.00

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
55 - 59.9	8	Beta Thalassaemia Major	1	0.16
		Beta Thalassaemia Intermedia	2	0.31
		HbE-Beta Thalassaemia	3	0.47
		HbH Disease	1	0.16
		Others*	1	0.16
60 - 64.9	3	Beta Thalassaemia Major	1	0.16
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	1	0.16
		HbH Disease	1	0.16
		Others*	0	0.00
Above 65	3	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.16
		HbE-Beta Thalassaemia	1	0.1
		HbH Disease	1	0.16
		Others*	0	0.00
Total			637	100.00

*Other types of alpha thalassaemia, excluding Hb Adana and HbH Constant Spring. Hb Adana and HbH Constant Spring are included in HbH disease.

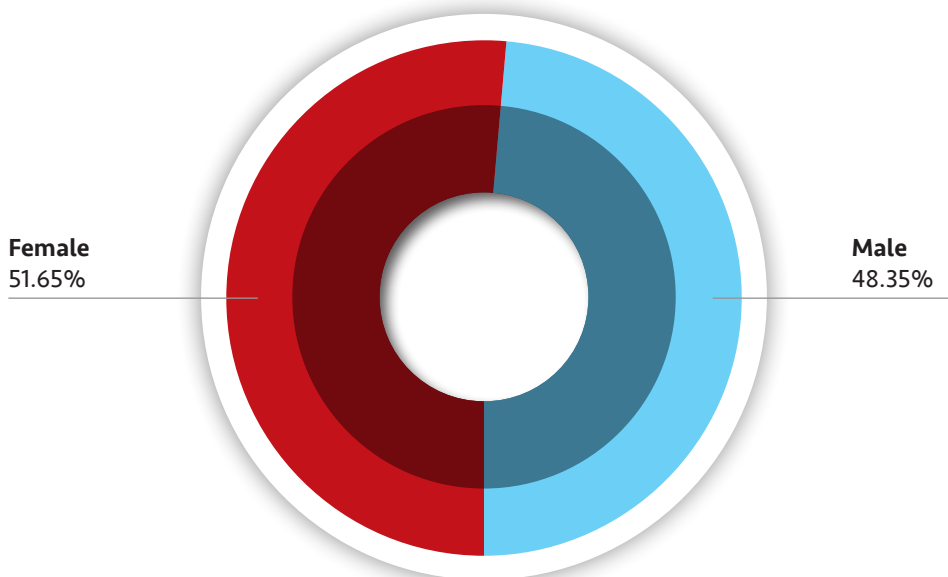
4.2.2 Gender

There are 308 male patients (48.35%) and 329 female patients (51.65%) registered in MTR in Johor. The distribution of male and female patients in each district hospital is as shown in Table 4.6.

Table 4.6: Distribution of Patients in Johor by Gender

Centre	Male		Female	
	No.	%	No.	%
Temenggung Seri Maharaja Tun Ibrahim, Kulaijaya	5	0.78	0	0.00
Sultanah Aminah, Johor Bahru	142	22.29	167	26.22
Pakar Sultanah Fatimah, Muar	40	6.28	34	5.34
Sultanah Nora Ismail, Batu Pahat	20	3.14	12	1.88
Enche' Besar Hajjah Kalsom, Kluang	20	3.14	27	4.24
Segamat	16	2.51	23	3.61
Pontian	2	0.31	5	0.78
Kota Tinggi	2	0.31	2	0.31
Mersing	10	1.57	17	2.67
Tangkak	0	0.00	0	0.00
Sultan Ismail, Johor Bahru	51	8.01	42	6.59
Total	308	48.35	329	51.65

Figure 4.4: Distribution of Patients in Johor by Gender



4.2.3 Ethnic Group

Majority of thalassaemia patients are Malays (483 patients, 75.82%), followed by Chinese (114 patients, 17.90%) and Indians (11 patients, 1.73%). Other ethnicities such as Orang Asli, Kadazan-Dusun, pribumi Sabah and Sarawak contributes 29 (4.55%) patients.

Table 4.7: Distribution of Patients in Johor by Ethnicity

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	483	75.82
Chinese	114	17.90
Indian	11	1.73
Others	29	4.55
Total	637	100

Figure 4.5: Distribution of Patients in Johor by Ethnic Group

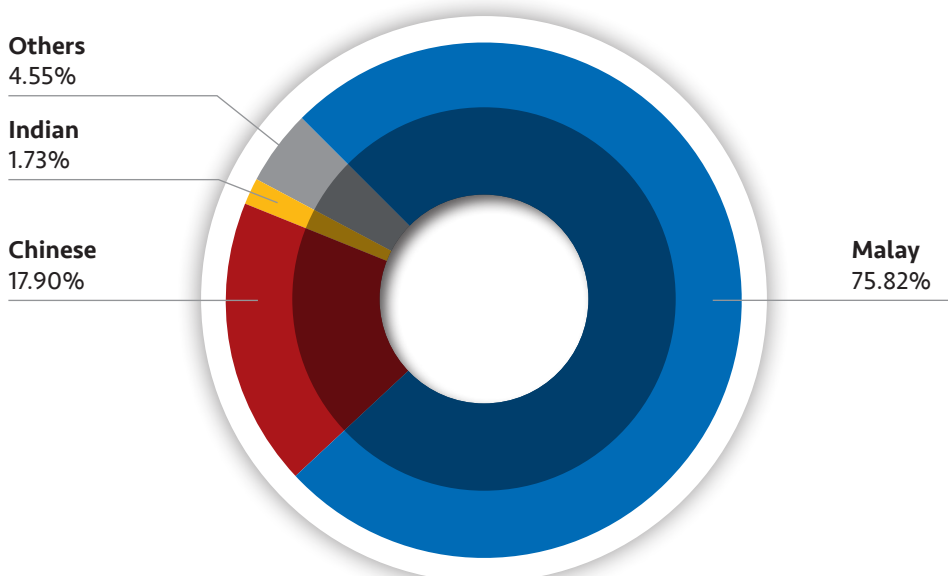
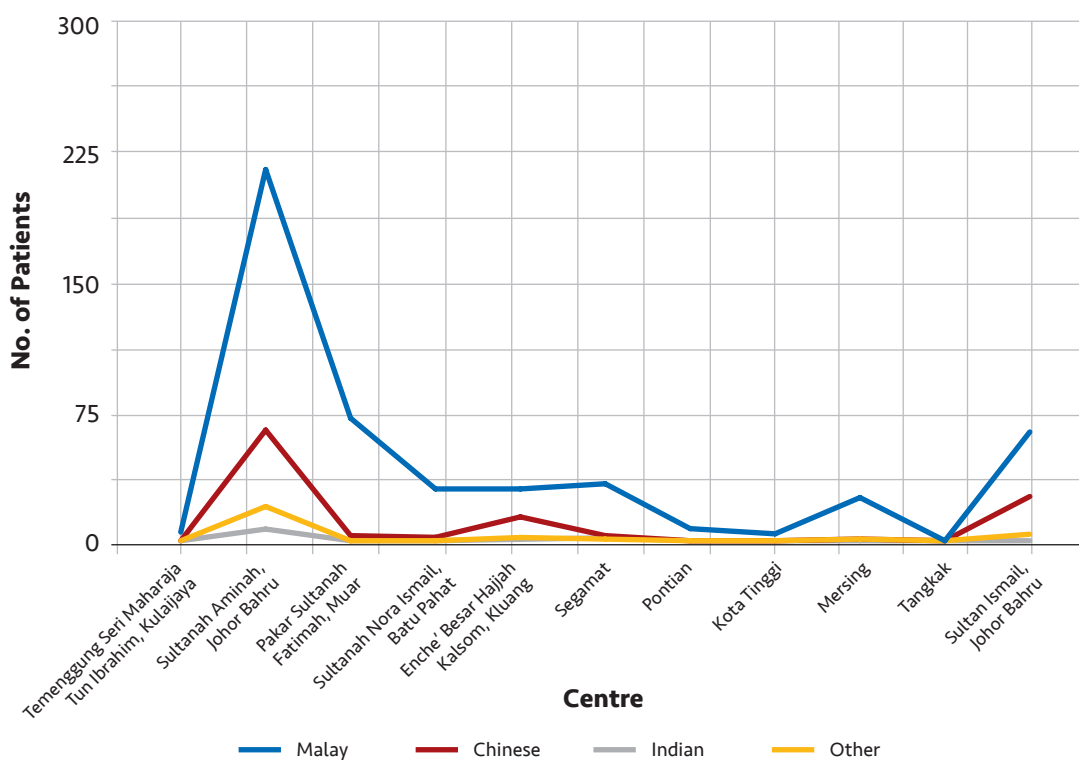


Table 4.8: Distribution of Patients in Johor According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Orang Asli		Kadazan		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Temenggung Seri Maharaja Tun Ibrahim, Kulajaya	5	0.78	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Sultanah Aminah, Johor Bahru	215	33.75	65	10.20	8	1.26	1	0.16	10	1.57	10	1.57
Pakar Sultanah Fatimah, Muar	71	11.15	3	0.47	0	0.00	0	0.00	0	0.00	0	0.00
Sultanah Nora Ismail, Batu Pahat	30	4.71	2	0.31	0	0.00	0	0.00	0	0.00	0	0.00
Enche' Besar Hajjah Kalsom, Kluang	30	4.71	14	2.20	1	0.16	0	0.00	2	0.31	0	0.00
Segamat	33	5.18	3	0.47	2	0.31	0	0.00	0	0.00	1	0.16
Pontian	7	1.10	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Kota Tinggi	4	0.63	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Mersing	25	3.92	1	0.16	0	0.00	1	0.16	0	0.00	0	0.00
Tangkak	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Sultan Ismail, Johor Bahru	63	9.89	26	4.08	0	0.00	0	0.00	3	0.47	1	0.16
Total	483	75.82	114	17.90	11	1.73	2	0.31	15	2.35	12	1.88

Figure 4.6: Distribution of Patients in Johor According to Ethnic Group by Centre



4.3 Diagnosis

Beta thalassaemia has the highest number of patients, with HbE-beta thalassaemia afflicting 264 patients (41.44%) followed by beta thalassaemia major (210 patients, 32.97%) and beta thalassaemia intermedia (53 patients, 8.32%). For alpha thalassaemia, 106 patients out of 110 patients (16.64%) have HbH disease and the remaining 4 patients (0.63%) have other types of alpha thalassaemia (excluding Hb Constant Spring and Hb Adana).

Table 4.9: Distribution of Patients in Johor by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	210	32.97
Beta Thalassaemia Intermedia	53	8.32
HbE-Beta Thalassaemia	264	41.44
HbH Disease	106	16.64
Others	4	0.63
Total	637	100

Figure 4.7: Distribution of Patients in Johor by Diagnosis

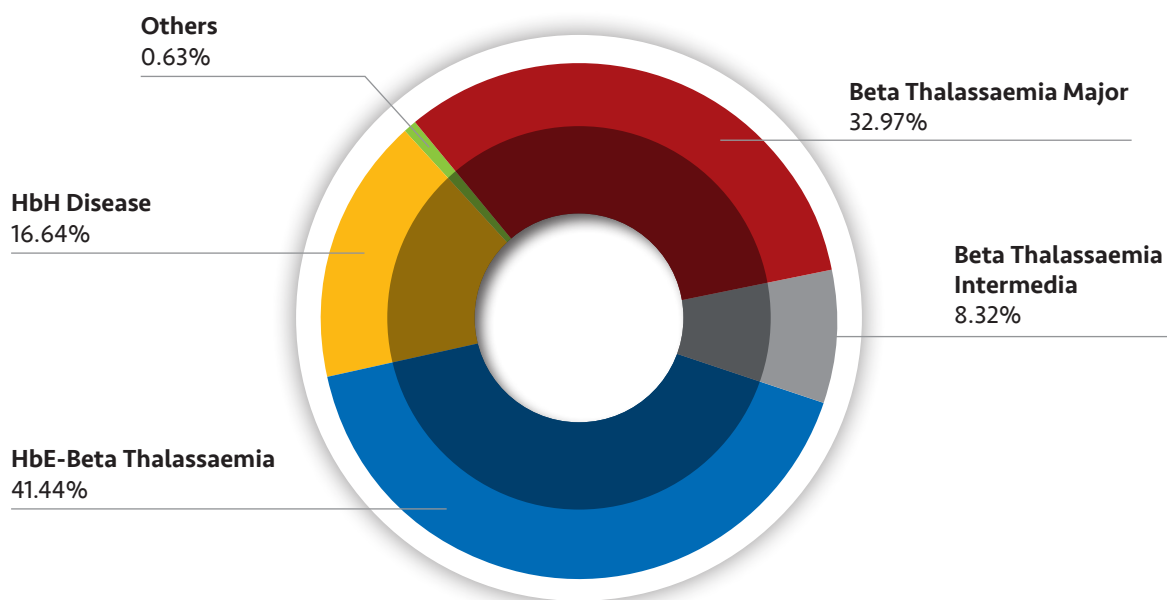


Table 4.10: Distribution of Patients in Johor According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Temenggung Seri Maharaja Tun Ibrahim, Kulajaya	4	0.63	0	0.00	0	0.00	1	0.16	0	0.00
Sultanah Aminah, Johor Bahru	88	13.81	33	5.18	123	19.31	64	10.05	1	0.16
Pakar Sultanah Fatimah, Muar	27	4.24	5	0.78	37	5.81	5	0.78	0	0.00
Sultanah Nora Ismail, Batu Pahat	7	1.10	2	0.31	21	3.30	2	0.31	0	0.00
Enche' Besar Hajjah Kalsom, Kluang	26	4.08	5	0.78	11	1.73	4	0.63	1	0.16
Segamat	14	2.20	4	0.63	11	1.73	9	1.41	1	0.16
Pontian	4	0.63	0	0.00	3	0.47	0	0.00	0	0.00
Kota Tinggi	2	0.31	1	0.16	1	0.16	0	0.00	0	0.00
Mersing	7	1.10	0	0.00	18	2.83	2	0.31	0	0.00
Tangkak	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Sultan Ismail, Johor Bahru	31	4.87	3	0.47	39	6.12	19	2.98	1	0.16
Total	210	32.97	53	8.32	264	41.44	106	16.64	4	0.63

Figure 4.8: Distribution of Patients in Johor According to Type of Thalassaemia by Age Group

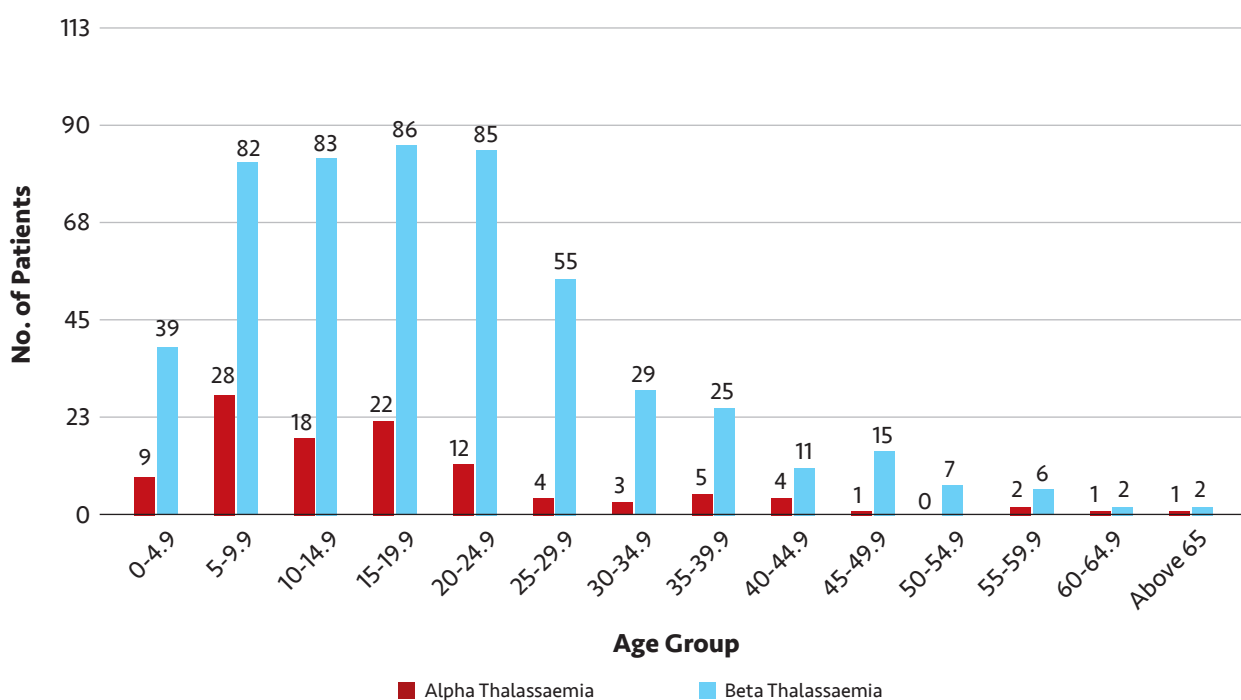


Table 4.11: Distribution of Patients in Johor According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	210	Malay	132	20.72
		Chinese	54	8.48
		Indian	6	0.94
		Kadazan-Dusun	11	1.73
		Orang Asli	1	0.16
		Others	6	0.94
Beta Thalassaemia Intermedia	53	Malay	39	6.12
		Chinese	7	1.10
		Indian	3	0.47
		Kadazan-Dusun	2	0.31
		Orang Asli	0	0.00
		Others	2	0.31
HbE-Beta Thalassaemia	264	Malay	248	38.93
		Chinese	15	2.35
		Indian	0	0.00
		Kadazan-Dusun	1	0.16
		Orang Asli	0	0.00
		Others	0	0.00
HbH Disease	106	Malay	62	9.73
		Chinese	36	5.65
		Indian	2	0.31
		Kadazan-Dusun	1	0.16
		Orang Asli	1	0.16
		Others	4	0.63
Others	4	Malay	2	0.31
		Chinese	2	0.31
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Orang Asli	0	0.00
		Others	0	0.00
Total			637	100.00

4.4 Treatment

4.4.1 Iron Chelation Therapy

A total of 422 patients (66.25%) are on iron chelation therapy. About 295 patients (46.31%) are on monotherapy, in which DFP contributes the highest number of patients followed by DFX and DFO. About 125 patients are receiving a two-drug combination and only two patients are on a three-drug combination. Combination of DFO and DFP is the more common practice (Figure 4.9). About 360 patients (91%) with TDT and 62 patients (26%) with non-transfusion-dependent thalassaemia (NTDT) require iron chelation therapy. The youngest NTDT patient receiving iron chelation therapy is 4 years old. Based on Figure 4.10, the most common iron chelator used by patients below the age of 15 is DFX.

Another 215 patients do not receive iron chelation therapy as they are from the NTDT group. A minority of these patients (8 patients) are either below 2 years old or has a serum ferritin level lower than 1000 ng/mL. This small group of patients may soon require iron chelation therapy. Distribution of patients receiving iron chelation therapy by type of iron chelator, centre and age group are shown in Tables 4.12-4.14 and Figures 4.9 and 4.10.

Table 4.12: Distribution of Patients in Johor by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	64	15.17
DFP only	119	28.20
DFX only	112	26.54
DFO + DFP	100	23.70
DFP + DFX	8	1.90
DFO + DFX	17	4.03
DFO + DFP + DFX	2	0.47
Total	422	100.00

Figure 4.9: Distribution of Patients in Johor by Type of Iron Chelator Received

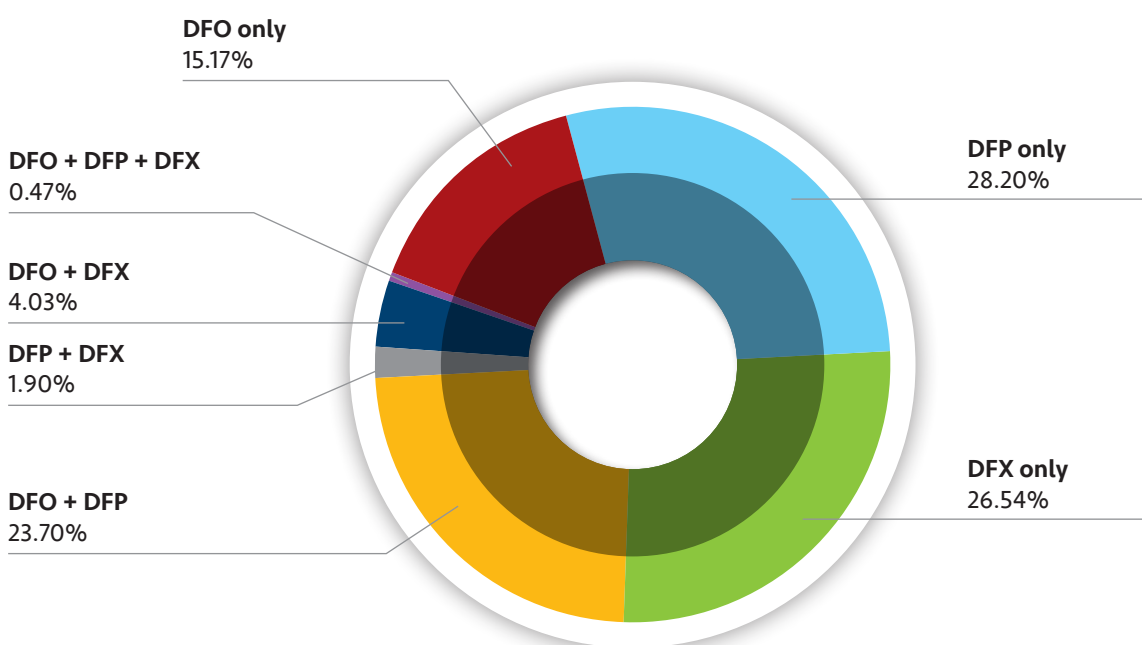


Table 4.13: Distribution of Patients in Johor According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Temenggung Seri Maharaja Tun Ibrahim, Kulai	4	DFO only	0	0.00
		DFP only	1	0.24
		DFX only	2	0.47
		DFO + DFP	1	0.24
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Sultanah Aminah, Johor Bahru	182	DFO only	31	7.35
		DFP only	65	15.40
		DFX only	21	4.98
		DFO + DFP	52	12.32
		DFP + DFX	4	0.95
		DFO + DFX	7	1.66
		DFO + DFP + DFX	2	0.47
Pakar Sultanah Fatimah, Muar	58	DFO only	14	3.32
		DFP only	8	1.90
		DFX only	13	3.08
		DFO + DFP	19	4.50
		DFP + DFX	2	0.47
		DFO + DFX	2	0.47
		DFO + DFP + DFX	0	0.00
Sultanah Nora Ismail, Batu Pahat	27	DFO only	4	0.95
		DFP only	3	0.71
		DFX only	12	2.84
		DFO + DFP	7	1.66
		DFP + DFX	0	0.00
		DFO + DFX	1	0.24
		DFO + DFP + DFX	0	0.00
Enche' Besar Hajjah Kalsom, Kluang	43	DFO only	10	2.37
		DFP only	7	1.66
		DFX only	11	2.61
		DFO + DFP	11	2.61
		DFP + DFX	1	0.24
		DFO + DFX	3	0.71
		DFO + DFP + DFX	0	0.00

4 Johor

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Segamat	25	DFO only	1	0.24
		DFP only	11	2.61
		DFX only	10	2.37
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	3	0.71
		DFO + DFP + DFX	0	0.00
Pontian	4	DFO only	0	0.00
		DFP only	3	0.71
		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
Kota Tinggi	2	DFO only	2	0.47
		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
Mersing	20	DFO only	0	0.00
		DFP only	10	2.37
		DFX only	4	0.95
		DFO + DFP	6	1.42
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Tangkak	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

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Sultan Ismail, Johor Bahru	57	DFO only	2	0.47
		DFP only	11	2.61
		DFX only	39	9.24
		DFO + DFP	3	0.71
		DFP + DFX	1	0.24
		DFO + DFX	1	0.24
		DFO + DFP + DFX	0	0.00
Total			422	100.00

Table 4.14: Distribution of Patients in Johor According to Type of Iron Chelator Received by Age Group

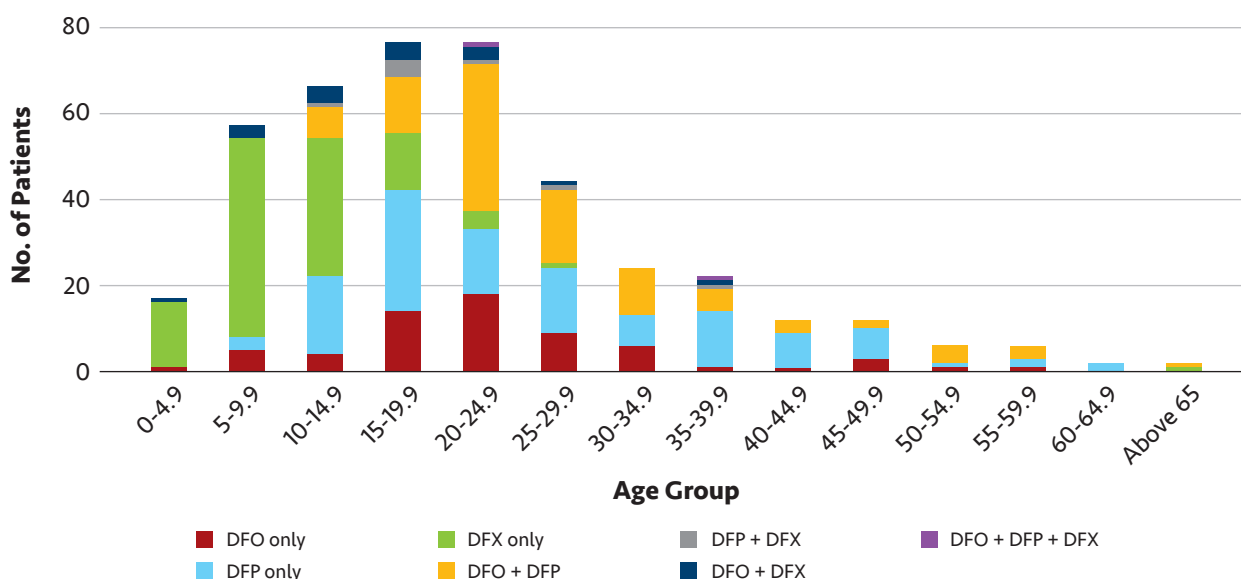
Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0 - 4.9	17	DFO only	1	0.24
		DFP only	0	0.00
		DFX only	15	3.55
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.24
		DFO + DFP + DFX	0	0.00
5 - 9.9	57	DFO only	5	1.18
		DFP only	3	0.71
		DFX only	46	10.90
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	3	0.71
		DFO + DFP + DFX	0	0.00
10 -14.9	66	DFO only	4	0.95
		DFP only	18	4.27
		DFX only	32	7.58
		DFO + DFP	7	1.66
		DFP + DFX	1	0.24
		DFO + DFX	4	0.95
		DFO + DFP + DFX	0	0.00
15 -19.9	76	DFO only	14	3.32
		DFP only	28	6.64
		DFX only	13	3.08
		DFO + DFP	13	3.08
		DFP + DFX	4	0.95
		DFO + DFX	4	0.95
		DFO + DFP + DFX	0	0.00

4 Johor

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
20 - 24.9	76	DFO only	18	4.27
		DFP only	15	3.55
		DFX only	4	0.95
		DFO + DFP	34	8.06
		DFP + DFX	1	0.24
		DFO + DFX	3	0.71
		DFO + DFP + DFX	1	0.24
25 - 29.9	44	DFO only	9	2.13
		DFP only	15	3.55
		DFX only	1	0.24
		DFO + DFP	17	4.03
		DFP + DFX	1	0.24
		DFO + DFX	1	0.24
		DFO + DFP + DFX	0	0.00
30 - 34.9	24	DFO only	6	1.42
		DFP only	7	1.66
		DFX only	0	0.00
		DFO + DFP	11	2.61
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
35 - 39.9	22	DFO only	1	0.24
		DFP only	13	3.08
		DFX only	0	0.00
		DFO + DFP	5	1.18
		DFP + DFX	1	0.24
		DFO + DFX	1	0.24
		DFO + DFP + DFX	1	0.24
40 - 44.9	12	DFO only	1	0.24
		DFP only	8	1.90
		DFX only	0	0.00
		DFO + DFP	3	0.71
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
45 - 49.9	12	DFO only	3	0.71
		DFP only	7	1.66
		DFX only	0	0.00
		DFO + DFP	2	0.47
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	6	DFO only	1	0.24
		DFP only	1	0.24
		DFX only	0	0.00
		DFO + DFP	4	0.95
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	6	DFO only	1	0.24
		DFP only	2	0.47
		DFX only	0	0.00
		DFO + DFP	3	0.71
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	2	DFO only	0	0.00
		DFP only	2	0.47
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	2	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	0.24
		DFO + DFP	1	0.24
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			422	100.00

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



4.4.2 Serum Ferritin Level

A total of 326 of TDT patients had their serum ferritin level measured in 2018. Of these, 43.87% has a serum ferritin level lower than 2500 ng/mL. Fifty-three patients (16.26%) have a serum ferritin level below 1000 ng/mL and 90 patients (27.61%) have a serum ferritin level between 1000-2499 ng/mL. However, 75 patients (23%) have a severe iron overload with serum ferritin level above than 5000 ng/mL. Most of the patients with high ferritin level are adolescents and adults who are not compliant to iron chelating agents. About 44% of regular follow-up patients did not have their serum ferritin level recorded in 2018. This can be due to poor record-keeping, such as a missing report from the patient’s record.

Table 4.15: Distribution of Patients in Johor by Most Recent Serum Ferritin Level

Serum Ferritin Level (ng/mL)	Centre	Total	< 1000		1000-2499		2500-4999		5000-9999		10,000+	
			No.	%	No.	%	No.	%	No.	%	No.	%
	Temengging Seri Maharaja Tun Ibrahim, Kulai	3	0	0.00	1	0.31	0	0.00	2	0.61	0	0.00
	Sultanah Aminah, Johor Bahru	131	30	9.20	36	11.04	45	13.80	17	5.21	3	0.92
	Pakar Sultanah Fatimah, Muar	46	5	1.53	13	3.99	15	4.60	12	3.68	1	0.31
	Sultanah Nora Ismail, Batu Pahat	23	0	0.00	3	0.92	13	3.99	7	2.15	0	0.00
	Enche' Besar Hajjah Kalsom, Kluang	26	3	0.92	8	2.45	8	2.45	6	1.84	1	0.31
	Segamat	22	3	0.92	3	0.92	6	1.84	4	1.23	6	1.84
	Pontian	1	0	0.00	0	0.00	1	0.31	0	0.00	0	0.00
	Kota Tinggi	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
	Mersing	20	4	1.23	2	0.61	6	1.84	8	2.45	0	0.00
	Tangkak	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
	Sultan Ismail, Johor Bahru	54	8	2.45	24	7.36	14	4.29	8	2.45	0	0.00
Total		326	53	16.26	90	27.61	108	33.13	64	19.63	11	3.37

4.5 Observation and Comments

1. The data needs to be updated and verified with the treating doctor, especially data for the adult patients.
2. Most patients with high serum ferritin level have poor adherence to iron chelation therapy, a non-functioning DFO pump, experience severe side effects or is unable to tolerate alternative iron chelators.
3. The percentage of patients with serum ferritin level recorded is low due to poor documentation and irregular monitoring. This may be due to budget constraints.
4. Most patients from Johor are in the Johor Bahru district due to the high population in the area and presence of two main haematology centres (Hospital Sultanah Aminah and Hospital Sultan Ismail).
5. Most patients aged 15 years and below are on DFX whereas a majority of adult patients are on DFP or drug combination therapy to reduce their serum ferritin level.
6. New thalassaemia birth cases is decreasing in trend. Most of new births were diagnosed as beta thalassaemia.

4.5.1 Recommendations

1. Closer working relationship between RAs and clinicians/paramedics.
2. RAs must trace the necessary information required in the registry.
3. TDT and NTD in the registry should also be included in the transfusion summary.
4. Each hospital must assign one paramedic to capture the latest data on thalassaemia patients, especially for adult patients.
5. Each patient in the registry should have their MRI T2* data recorded.
6. To add on free entry write in annual comment section for clinicians.
7. To increase financial budget for iron chelation therapy, monitoring test for patients, blood filters as well as payment for RA.

5 Kedah

5.1. Introduction

Kedah has an estimated population of 2,163,700 people in 2018 (Department of Statistics Malaysia). Majority of thalassaemia patients in Kedah are receiving care at Hospital Sultanah Bahiyah, Hospital Sultan Abdul Halim and Hospital Kulim.

5.2. Patient Demographics

There is a total of 626 living patients in Kedah. Two patients were cured by stem cell transplant and 31 patients have succumbed to the disease. The distribution of patients in Kedah is shown in Table 5.1.

Table 5.1: Distribution of Patients in Kedah by Centre

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
Hospital Sultanah Bahiyah, Alor Setar	358	51.59
Hospital Baling	13	1.87
Hospital Jitra	0	0.00
Hospital Kuala Nerang	3	0.43
Hospital Kulim	127	18.30
Hospital Langkawi	48	6.92
Hospital Sik	10	1.44
Hospital Sultan Abdul Halim, Sungai Petani	135	19.45
Hospital Yan	0	0.00
Total	694	100

Figure 5.1: Distribution of Patients in Kedah by Centre

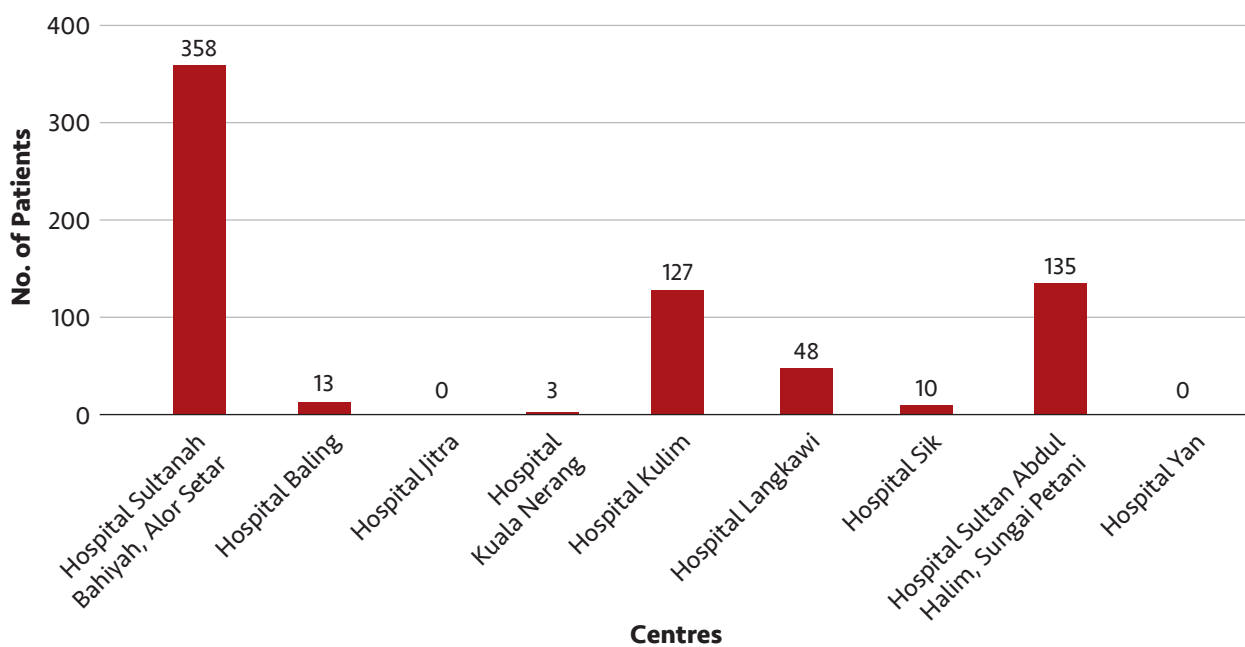




Table 5.2: Distribution of Patients in Kedah by Vital Status

Vital Status	No. Of Patients
Alive	626
Cumulative Cured by Stem Cell Therapy	2
Lost to Follow-up	66
Total	694
Deaths in 2018	3
Cumulative Reported Deaths	31

Table 5.3: Cumulative Causes of Death Since 2007 in Kedah

Cause of Death	No. of Patients
Infections	23
Cardiac Causes	1
Intracranial Bleed	1
Liver Disease (Hepatic Failure)	1
Endocrine Complication (Renal Failure)	1
Motor Vehicle Accident	2
Severe Head Injury	1
Stomach Ache	1
Total	31

5.2.1 Age

As shown in Table 5.4, the distribution of patients in Kedah is dominated by paediatric patients. A total of 451 patients (64.99%) are between 0-19.9 years old. The youngest patient in Kedah is 1 year old and the eldest is 70 years old.

Table 5.4: Distribution of Patients in Kedah by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 – 4.9	22	3.17
5 – 9.9	130	18.73
10 – 14.9	154	22.19
15 – 19.9	145	20.89
20 – 24.9	122	17.58
25 – 29.9	64	9.22
30 – 34.9	28	4.03
35 – 39.9	7	1.01
40 – 44.9	8	1.15
45 – 49.9	7	1.01
50 – 54.9	3	0.43
55 – 59.9	1	0.14
60 – 64.9	1	0.14
Above 65	2	0.29
Total	694	100.00

Figure 5.2: Distribution of Patients in Kedah by Age Group

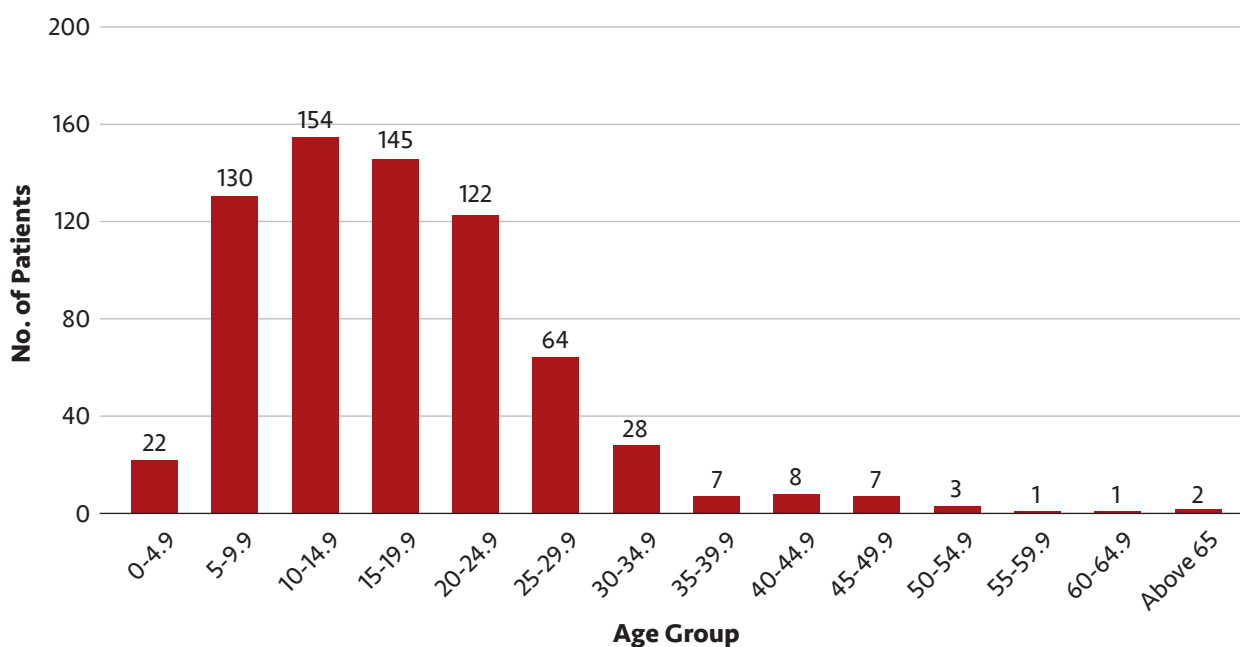


Table 5.5: Distribution of Patients in Kedah According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 – 4.9	22	Beta Thalassaemia Major	4	0.58
		Beta Thalassaemia Intermedia	4	0.58
		HbE-Beta Thalassaemia	8	1.15
		HbH Disease	6	0.86
		Others	0	0.00
5 – 9.9	130	Beta Thalassaemia Major	19	2.74
		Beta Thalassaemia Intermedia	8	1.15
		HbE-Beta Thalassaemia	48	6.92
		HbH Disease	40	5.76
		Others	15	2.16
10 – 14.9	154	Beta Thalassaemia Major	17	2.45
		Beta Thalassaemia Intermedia	9	1.30
		HbE-Beta Thalassaemia	71	10.23
		HbH Disease	39	5.62
		Others	18	2.59
15 – 19.9	145	Beta Thalassaemia Major	22	3.17
		Beta Thalassaemia Intermedia	9	1.30
		HbE-Beta Thalassaemia	66	9.51
		HbH Disease	38	5.48
		Others	10	1.44
20 – 24.9	122	Beta Thalassaemia Major	19	2.74
		Beta Thalassaemia Intermedia	7	1.01
		HbE-Beta Thalassaemia	52	7.49
		HbH Disease	33	4.76
		Others	11	1.59
25 – 29.9	64	Beta Thalassaemia Major	17	2.45
		Beta Thalassaemia Intermedia	4	0.58
		HbE-Beta Thalassaemia	26	3.75
		HbH Disease	16	2.31
		Others	1	0.14
30 – 34.9	28	Beta Thalassaemia Major	7	1.01
		Beta Thalassaemia Intermedia	3	0.43
		HbE-Beta Thalassaemia	13	1.87
		HbH Disease	5	0.72
		Others	0	0.00

5 Kedah

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	7	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.14
		HbE-Beta Thalassaemia	3	0.43
		HbH Disease	2	0.29
		Others	1	0.14
40 - 44.9	8	Beta Thalassaemia Major	1	0.14
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	2	0.29
		HbH Disease	4	0.58
		Others	1	0.14
45 - 49.9	7	Beta Thalassaemia Major	1	0.14
		Beta Thalassaemia Intermedia	2	0.29
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	3	0.43
		Others	1	0.14
50 - 54.9	3	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	0.29
		HbE-Beta Thalassaemia	1	0.14
		HbH Disease	0	0.00
		Others	0	0.00
55 - 59.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.14
		Others	0	0.00
60 - 64.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	1	0.14
Above 65	2	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.14
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.14
		Others	0	0.00
Total			694	100

5.2.2 Gender

There are 370 male patients (53.31%) and 324 female patients (46.69%) registered in MTR in Kedah. The distribution of the male and female patients in each district hospital is as shown in Table 5.6 and Figure 5.3.

Figure 5.3: Distribution of Patients in Kedah by Gender

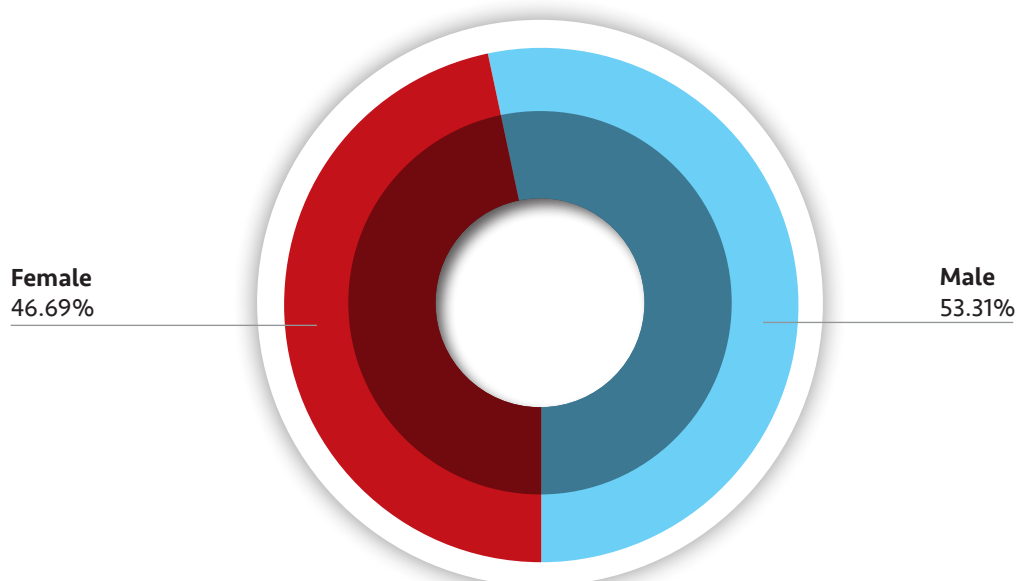


Table 5.6: Distribution of Patients in Kedah by Gender

Centre	Male		Female	
	No.	%	No.	%
Hospital Sultanah Bahiyah, Alor Setar	180	25.94	178	25.65
Hospital Baling	8	1.15	5	0.72
Hospital Jitra	0	0.00	0	0.00
Hospital Kuala Nerang	0	0.00	3	0.43
Hospital Kulim	67	9.65	60	8.65
Hospital Langkawi	20	2.88	28	4.03
Hospital Sik	9	1.30	1	0.14
Hospital Sultan Abdul Halim, Sungai Petani	86	12.39	49	7.06
Hospital Yan	0	0.00	0	0.00
Total	370	53.31	324	46.69

5.2.3 Ethnic Group

The distribution of patients in Kedah by ethnic group is shown in Table 5.7. A large majority (654 patients, 94.24%) are Malay, 17 patients (2.45%) are Chinese and 2 patients (0.29%) are Indian. Another 21 patients (3.02%) are of other ethnicity. Table 5.8 shows the distribution of the patients according to ethnic group by centre in Kedah.

Table 5.7: Distribution of Patients in Kedah by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	654	94.24
Chinese	17	2.45
Indian	2	0.29
Others	21	3.02
Total	694	100

Figure 5.4: Distribution of Patients in Kedah by Ethnic Group

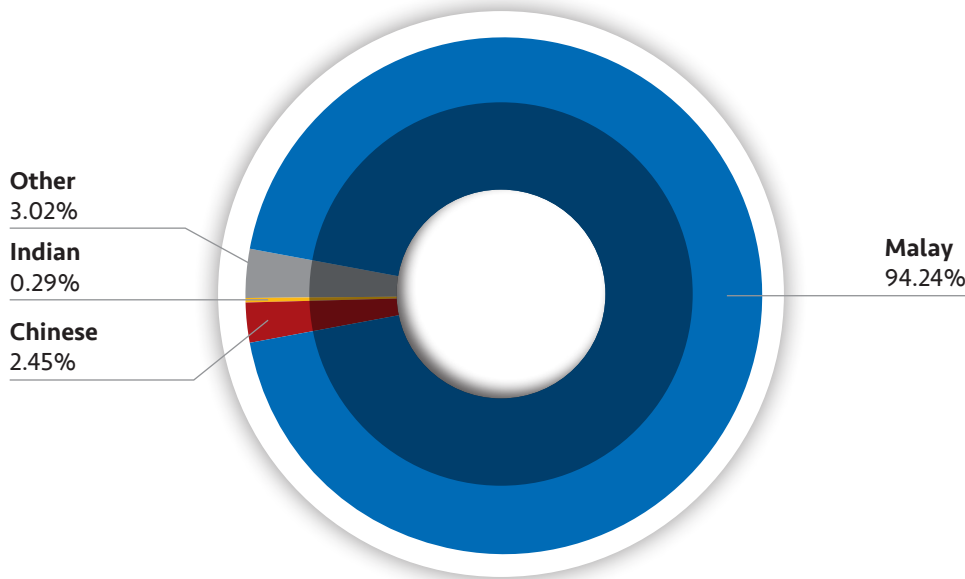


Table 5.8: Distribution of Patients in Kedah According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Thai		Mixed		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Bahiyah, Alor Setar	331	47.69	10	1.44	1	0.14	14	2.02	0	0.00	2	0.29
Hospital Baling	13	1.87	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Jitra	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kuala Nerang	2	0.29	0	0.00	0	0.00	1	0.14	0	0.00	0	0.00
Hospital Kulim	123	17.72	2	0.29	1	0.14	0	0.00	0	0.00	1	0.14
Hospital Langkawi	47	6.77	1	0.14	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Sik	9	1.30	0	0.00	0	0.00	1	0.14	0	0.00	0	0.00
Hospital Sultan Abdul Halim, Sungai Petani	129	18.59	4	0.58	0	0.00	2	0.29	0	0.00	0	0.00
Hospital Yan	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	654	94.24	17	2.45	2	0.29	18	2.59	0	0	3	0.43

5.3. Diagnosis

HbE-beta thalassaemia has the highest number of patients (290, 41.79%), followed by HbH disease (188 patients, 27.09%), beta thalassaemia major (107 patients, 15.42%), other diagnosis (59 patients, 8.50%) and beta thalassaemia intermedia (50 patients, 7.20%).

Table 5.9: Distribution of Patients in Kedah by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	107	15.42
Beta Thalassaemia Intermedia	50	7.20
HbE-Beta Thalassaemia	290	41.79
HbH Disease	188	27.09
Others	59	8.50
Total	694	100

5 Kedah

Figure 5.5: Distribution of Patients in Kedah by Diagnosis

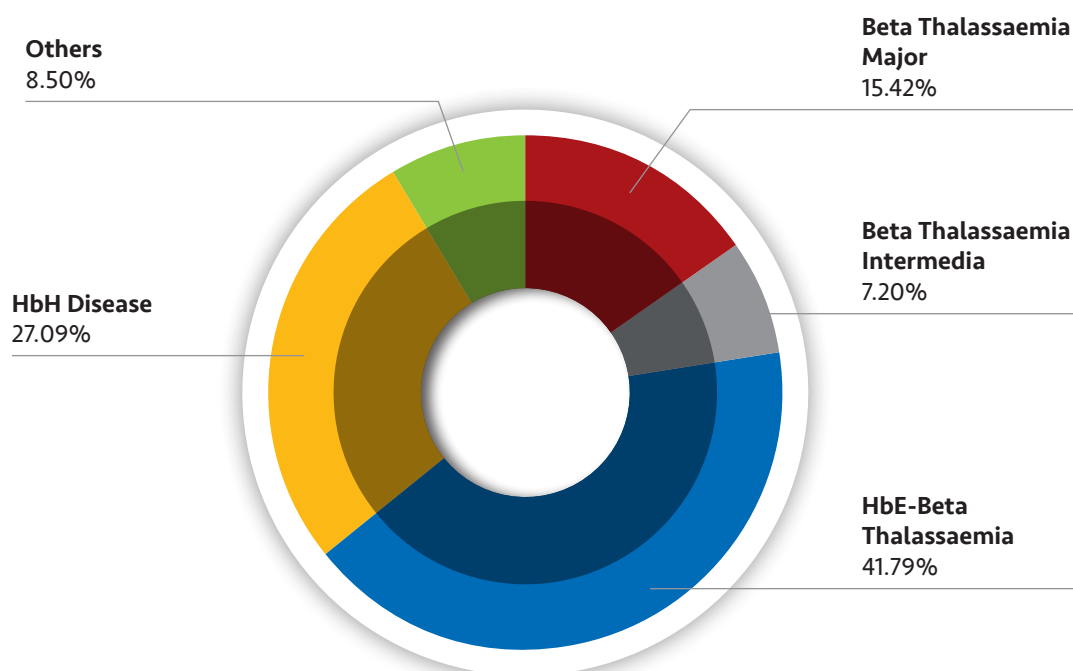


Table 5.10: Distribution of Patients in Kedah According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Sultanah Bahiyah, Alor Setar	66	9.51	21	3.03	147	21.18	97	13.98	27	3.89
Hospital Baling	3	0.43	0	0.00	7	1.01	3	0.43	0	0.00
Hospital Jitra	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kuala Nerang	0	0.00	0	0.00	1	0.14	0	0.00	2	0.29
Hospital Kulim	11	1.59	14	2.02	54	7.78	34	4.90	14	2.02
Hospital Langkawi	3	0.43	5	0.72	16	2.31	18	2.59	6	0.86
Hospital Sik	1	0.14	0	0.00	8	1.15	1	0.14	0	0.00
Hospital Sultan Abdul Halim, Sungai Petani	23	3.31	10	1.44	57	8.21	35	5.04	10	1.44
Hospital Yan	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	107	15.42	50	7.20	290	41.79	188	27.09	59	8.50

Table 5.11: Distribution of Patients in Kedah According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	107	Malay	98	14.12
		Chinese	4	0.58
		Indian	1	0.14
		Others	4	0.58
Beta Thalassaemia Intermedia	50	Malay	48	6.92
		Chinese	2	0.29
		Indian	0	0.00
		Others	0	0.00
HbE-Beta Thalassaemia	290	Malay	278	40.06
		Chinese	8	1.15
		Indian	1	0.14
		Others	3	0.43
HbH Disease	188	Malay	173	24.93
		Chinese	3	0.43
		Indian	0	0.00
		Others	12	1.73
Others	59	Malay	57	8.21
		Chinese	0	0.00
		Indian	0	0.00
		Others	2	0.29
Total			694	100.00

5.4 Treatment

5.4.1 Iron Chelation

As shown in Table 5.12, out of 371 chelated patients, 53.91% are on oral DFX, 17.51% are on combination therapy, 9.70% are on subcutaneous DFO and 18.87% are on oral DFP.

Table 5.12: Distribution of Patients in Kedah by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	36	9.70
DFP only	70	18.87
DFX only	200	53.91
DFO + DFP	42	11.32
DFP + DFX	11	2.96
DFO + DFX	11	2.96
DFO + DFP + DFX	1	0.27
Total	371	100.00

Table 5.13: Distribution of Patients in Kedah According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Sultanah Bahiyah, Alor Setar	191	DFO only	7	1.89
		DFP only	39	10.51
		DFX only	108	29.11
		DFO + DFP	19	5.12
		DFP + DFX	10	2.70
		DFO + DFX	7	1.89
		DFO + DFP + DFX	1	0.27
Hospital Baling	7	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	6	1.62
		DFO + DFP	1	0.27
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Jitra	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kuala Nerang	2	DFO only	0	0.00
		DFP only	1	0.27
		DFX only	1	0.27
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kulim	66	DFO only	10	2.70
		DFP only	16	4.31
		DFX only	26	7.01
		DFO + DFP	12	3.23
		DFP + DFX	1	0.27
		DFO + DFX	1	0.27
		DFO + DFP + DFX	0	0.00

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Langkawi	24	DFO only	0	0.00
		DFP only	6	1.62
		DFX only	15	4.04
		DFO + DFP	1	0.27
		DFP + DFX	0	0.00
		DFO + DFX	2	0.54
		DFO + DFP + DFX	0	0.00
Hospital Sik	6	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	6	1.62
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Sultan Abdul Halim, Sungai Petani	75	DFO only	19	5.12
		DFP only	8	2.16
		DFX only	38	10.24
		DFO + DFP	9	2.43
		DFP + DFX	0	0.00
		DFO + DFX	1	0.27
		DFO + DFP + DFX	0	0.00
Hospital Yan	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			371	100.00

Figure 5.6: Distribution of Patients in Kedah by Type of Iron Chelator Received

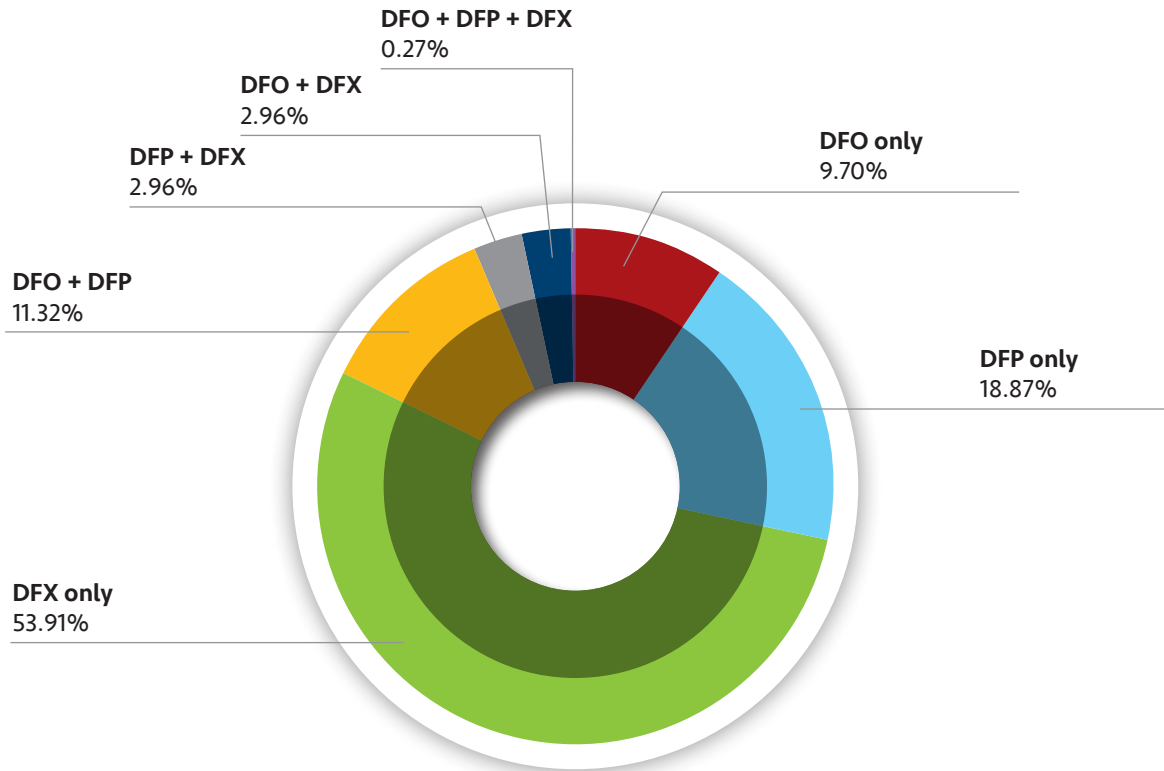


Table 5.14: Distribution of Patients in Kedah According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0 - 4.9	9	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	9	2.43
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	60	DFO only	0	0.00
		DFP only	1	0.27
		DFX only	58	15.63
		DFO + DFP	0	0.00
		DFP + DFX	1	0.27
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
10 -14.9	81	DFO only	4	1.08
		DFP only	6	1.62
		DFX only	64	17.25
		DFO + DFP	6	1.62
		DFP + DFX	0	0.00
		DFO + DFX	1	0.27
		DFO + DFP + DFX	0	0.00
15 -19.9	86	DFO only	9	2.43
		DFP only	16	4.31
		DFX only	45	12.13
		DFO + DFP	11	2.96
		DFP + DFX	3	0.81
		DFO + DFX	2	0.54
		DFO + DFP + DFX	0	0.00
20 - 24.9	60	DFO only	15	4.04
		DFP only	11	2.96
		DFX only	15	4.04
		DFO + DFP	10	2.70
		DFP + DFX	3	0.81
		DFO + DFX	6	1.62
		DFO + DFP + DFX	0	0.00
25 - 29.9	42	DFO only	6	1.62
		DFP only	19	5.12
		DFX only	4	1.08
		DFO + DFP	8	2.16
		DFP + DFX	3	0.81
		DFO + DFX	2	0.54
		DFO + DFP + DFX	0	0.00
30-34.9	21	DFO only	1	0.27
		DFP only	11	2.96
		DFX only	1	0.27
		DFO + DFP	7	1.89
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	0.27

5 Kedah

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
35-39.9	3	DFO only	0	0.00
		DFP only	3	0.81
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	2	DFO only	0	0.00
		DFP only	1	0.27
		DFX only	1	0.27
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-49.9	3	DFO only	0	0.00
		DFP only	1	0.27
		DFX only	1	0.27
		DFO + DFP	0	0.00
		DFP + DFX	1	0.27
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50-54.9	2	DFO only	1	0.27
		DFP only	1	0.27
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	0.27
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
60-64.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	0.27
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			371	100.00

5.4.2 Serum Ferritin Level

There are 266 patients in Kedah with their serum ferritin level recorded. There are 48 patients (18.05%) with serum ferritin level lower than 1000 ng/mL, 102 patients (38.35%) with serum ferritin level between 1000-2499 ng/mL, 71 patients (26.69%) with serum ferritin level between 2500-4999 ng/mL, and 45 patients (16.92%) with serum ferritin level above 5000 ng/mL.

Table 5.15: Distribution of Patients in Kedah by Most Recent Serum Ferritin Level

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Baling	6	0	0.00	2	0.75	3	1.13	0	0.00	1	0.38
Hospital Sultanah Bahiyah, Alor Setar	150	25	9.40	58	21.80	36	13.53	23	8.65	8	3.01
Hospital Kuala Nerang	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kulim	44	10	3.76	19	7.14	12	4.51	2	0.75	1	0.38
Hospital Langkawi	20	0	0.00	6	2.26	8	3.01	4	1.50	2	0.75
Hospital Sik	4	0	0.00	1	0.38	2	0.75	1	0.38	0	0.00
Hospital Sultan Abdul Halim, Sungai Petani	42	13	4.89	16	6.02	10	3.76	3	1.13	0	0.00
Hospital Yan	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	266	48	18.05	102	38.35	71	26.69	33	12.41	12	4.51

5.5 Observation and Comments

Nearly 65% of the cases in Kedah involves patients below 20 years old. HbE-beta thalassaemia is the most common type, afflicting 41.79% patients in Kedah. Most chelated patients (53.91%) are on oral DFX. Based on the serum ferritin level, 56.39% of thalassaemia patients have satisfactory iron chelation, with serum ferritin level < 2500 ng/mL.

The poor compliance among younger patients is due to poor supervision from parents. Iron chelation therapy should be optimised in these patients by ensuring continuous education to ensure better patient compliance. For those with high ferritin level, a combination therapy should be considered to reduce long term complications of iron overload. Some patients on subcutaneous DFO infusion also face difficulties in obtaining an infusion pump due to financial constraints.

Only Hospital Sultan Abdul Halim has dedicated day care facilities for thalassaemia patients in Kedah. As such, patients in Kedah face added financial burden to travel twice monthly for treatment.

Nearly 75% of deaths were caused by infections. However, we were not able to retrieve the details of causative organisms. In addition, 66 patients were lost to follow-up for unknown reasons.

5.5.1 Recommendations

- The Ministry of Health Malaysia should consider providing infusion pumps to patients who are on subcutaneous DFO.
- Medication Therapy Adherence Clinic (MTAC) pharmacists should be available in all thalassaemia clinics.
- All centres must work towards having a dedicated day care service for thalassaemia patients.
- Each centre must have a system to recall patients who default.
- It will be beneficial to document details on infectious causative organisms and comorbidities in patients who passed away.

5.5.2 Conclusion

Kedah has the fourth highest number of thalassaemia patients in Malaysia. The most common type of thalassaemia in Kedah is HbE-beta thalassaemia, followed by HbH disease. The median age group is 15-19.9 years old. Two hundred patients are on chelation therapy, with oral DFX being the most common chelator. Around 56.39% of patients achieved serum ferritin level below 2500 ng/mL. Poor adherence to treatment is mainly due to social and financial issues. A significant proportion of deaths are caused by infections.

6 Kelantan

6.1 Introduction

Kelantan is a state located in the northeast of Peninsular Malaysia. Kelantan's population is estimated at 1.86 million in 2018. The population is dominated by those of Malay descent, followed by the Chinese, Siamese and Indians. This state is managed by ten administrative jurisdictions, namely Kota Bharu, Kubang Kerian, Tumpat, Pasir Puteh, Pasir Mas, Tanah Merah, Machang, Kuala Krai, Jeli, and Gua Musang.

Kelantan has quite large number of thalassaemia patients who are seeking treatment. Thus, there are ten hospitals involved in providing treatment to thalassaemia 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 thalassaemia patients in Kelantan registered in the MTR are 486 patients. More than half of these patients received treatment at HRPZII and HUSM. In Kelantan, HRPZII, HUSM and Hospital Kuala Krai provided day care services including blood transfusions for thalassaemia patients.

6.2 Patient Demographics

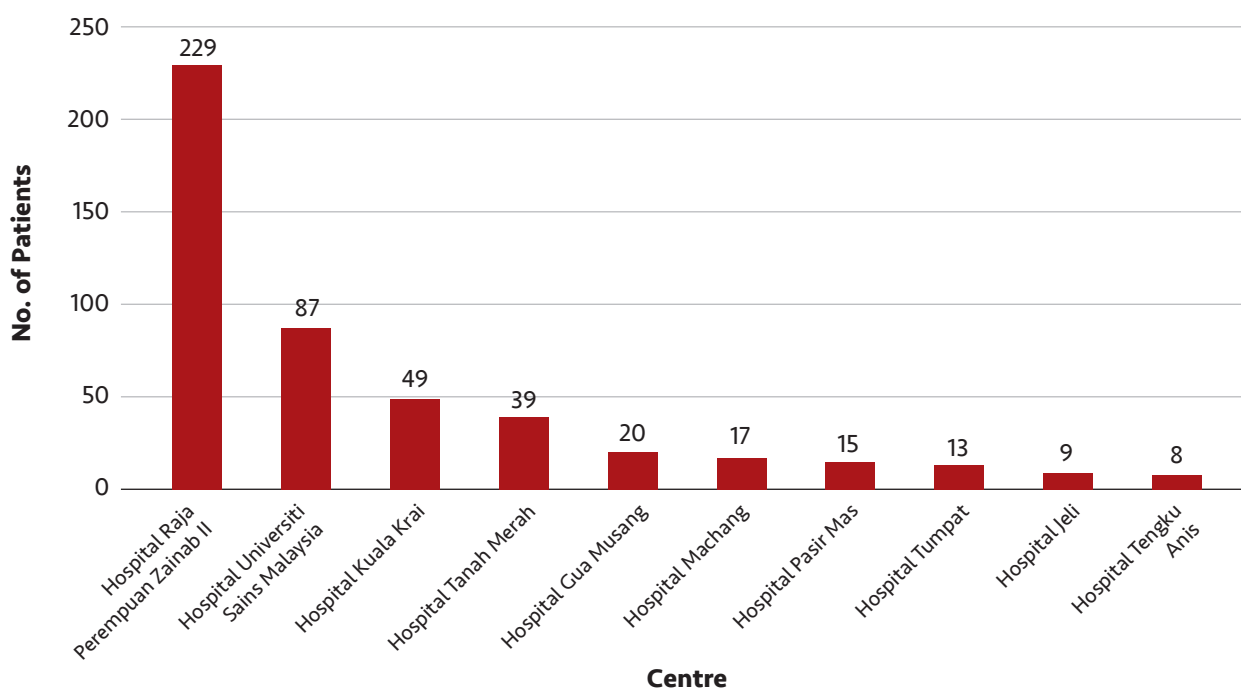
There are ten hospitals involved in providing treatment to thalassaemia patients in Kelantan. Table 6.1 shows the distribution of thalassaemia patients by centre in Kelantan. HRPZII has the greatest number of thalassaemia patients whereas Hospital Tengku Anis has the least.

Table 6.1: Distribution of Patients in Kelantan by Centre

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
Hospital Raja Perempuan Zainab II	229	47.12
Hospital Universiti Sains Malaysia	87	17.90
Hospital Kuala Krai	49	10.08
Hospital Tanah Merah	39	8.02
Hospital Gua Musang	20	4.12
Hospital Machang	17	3.50
Hospital Pasir Mas	15	3.09
Hospital Tumpat	13	2.67
Hospital Jeli	9	1.85
Hospital Tengku Anis	8	1.65
Total	486	100.00

6 Kelantan

Figure 6.1: Distribution of Patients in Kelantan by Centre



Only data of patients who are either alive, lost to follow-up or cured by bone marrow transplant are analysed. The total number of thalassaemia patients with any one of these three statuses is 486. In 2018, there was only one patient successfully cured by bone marrow transplant (BMT) in Kelantan.

Table 6.2: Distribution of Patients in Kelantan by Vital Status

Vital Status	Patients
Alive	428
Cumulative Reported Cured by Stem Cell Therapy	1
Lost to Follow-up	57
Total	486
Deaths in 2018	4
Cumulative Reported Deaths	29

In 2018, there were four deaths of thalassaemia patients recorded. Based on the report, two of the cases were due to cardiac causes and one was infection-related. The remaining one patient's cause of death was not able to be determined (patient died at home).

Table 6.3: Cumulative Causes of Death Since 2007 in Kelantan

Cause of Death	No. of Patients
Infection	11
Cardiac	11
Thrombosis	1
Unknown*	5
Others:	
Drowned	1
Total	29

*Missing data.

There were 29 deaths of thalassaemia patients since 2007 in Kelantan. Table 6.3 shows the causes of these deaths and number of patients for each cause of death. The most common causes of death among thalassaemia patients were infections and cardiac issues. There were five cases with unknown cause of death. Of the five, three patients' case notes were already disposed of by the medical record unit. The other two deceased patients' cause of death were not able to be determined.

6.2.1 Age

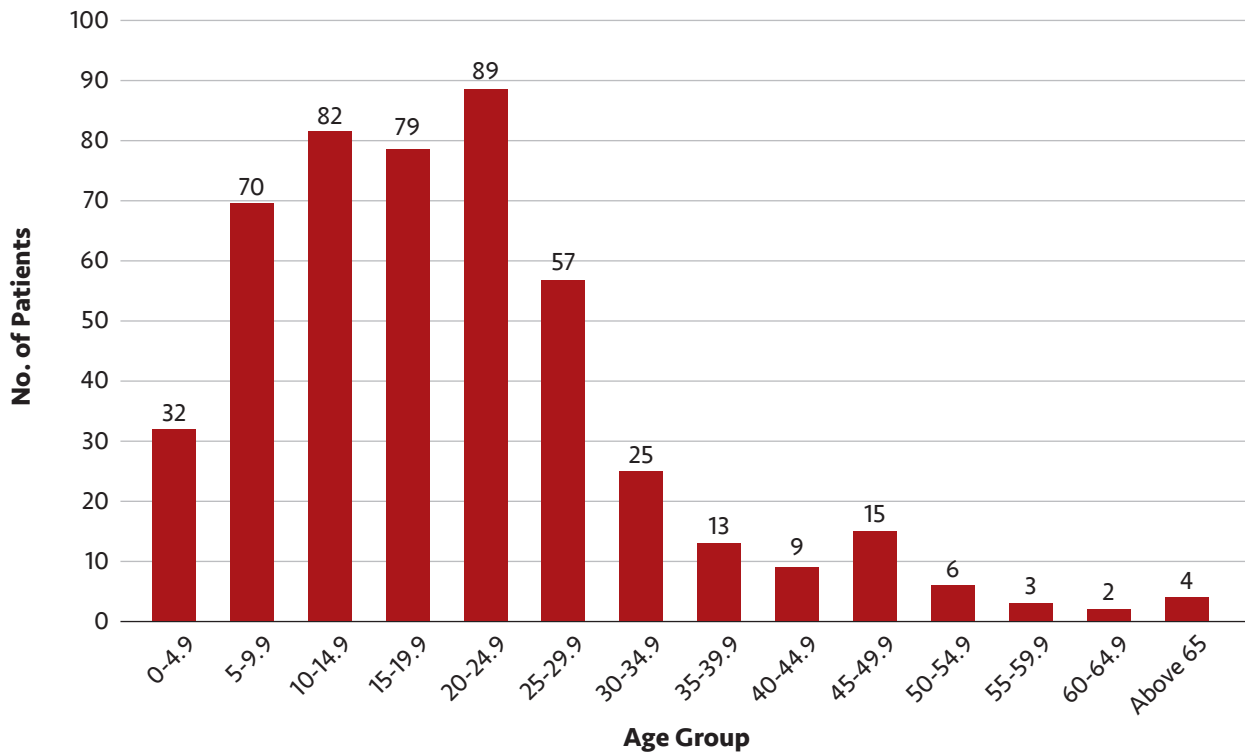
As shown in Table 6.4, most patients in Kelantan diagnosed with thalassaemia were young adults. Patients aged between 20-24.9 years leads with 89 patients (18.31%), followed patients aged between 10-14.9 with 82 patients (16.87%). The age range with the least number of thalassaemia patients were 60-64.9 years with 2 patients (0.41%).

Table 6.4: Distribution of Patients in Kelantan by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	32	6.58
5 - 9.9	70	14.40
10 - 14.9	82	16.87
15 - 19.9	79	16.26
20 - 24.9	89	18.31
25 - 29.9	57	11.73
30 - 34.9	25	5.14
35 - 39.9	13	2.67
40 - 44.9	9	1.85
45 - 49.9	15	3.09
50 - 54.9	6	1.23
55 - 59.9	3	0.62
60 - 64.9	2	0.41
Above 65	4	0.82
Total	486	100.0

6 Kelantan

Figure 6.2: Distribution of Patients in Kelantan by Age Group



The youngest patient (9 months of age) in Kelantan was diagnosed as beta thalassaemia major. On the other hand, the eldest beta thalassaemia major patient is 41 years old. There were four patients above 65 years old, the eldest of which is an 84-year-old diagnosed with alpha thalassaemia (presumably NTD).

Table 6.5: Distribution of Patients in Kelantan According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	32	Beta Thalassaemia Major	8	1.65
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	10	2.10
		HbH Disease	7	1.44
		Others	6	1.23
5 - 9.9	70	Beta Thalassaemia Major	11	2.26
		Beta Thalassaemia Intermedia	4	0.82
		HbE-Beta Thalassaemia	31	6.38
		HbH Disease	19	3.91
		Others	5	1.03
10 -14.9	82	Beta Thalassaemia Major	9	1.85
		Beta Thalassaemia Intermedia	8	1.65
		HbE-Beta Thalassaemia	36	7.40
		HbH Disease	23	4.73
		Others	6	1.23

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
15 -19.9	79	Beta Thalassaemia Major	11	2.26
		Beta Thalassaemia Intermedia	2	0.41
		HbE-Beta Thalassaemia	49	10.08
		HbH Disease	15	3.09
		Others	2	0.41
20 - 24.9	89	Beta Thalassaemia Major	11	2.26
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	52	10.70
		HbH Disease	24	4.94
		Others	1	0.21
25 - 29.9	57	Beta Thalassaemia Major	12	2.47
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	30	6.17
		HbH Disease	13	2.67
		Others	1	0.21
30 - 34.9	25	Beta Thalassaemia Major	4	0.82
		Beta Thalassaemia Intermedia	3	0.62
		HbE-Beta Thalassaemia	12	2.47
		HbH Disease	6	1.23
		Others	0	0.00
35 - 39.9	13	Beta Thalassaemia Major	3	0.62
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	6	1.23
		HbH Disease	2	0.41
		Others	1	0.21
40 - 44.9	9	Beta Thalassaemia Major	1	0.21
		Beta Thalassaemia Intermedia	2	0.41
		HbE-Beta Thalassaemia	5	1.03
		HbH Disease	1	0.21
		Others	0	0.00
45 - 49.9	15	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	4	0.82
		HbE-Beta Thalassaemia	6	1.23
		HbH Disease	4	0.82
		Others	1	0.21
50 - 54.9	6	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	1	0.21
		HbH Disease	1	0.21
		Others	3	0.62

6 Kelantan

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
55 - 59.9	3	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	1	0.21
		HbH Disease	1	0.21
		Others	0	0.00
60 - 64.9	2	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	1	0.21
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	4	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	1	0.21
		HbH Disease	1	0.21
		Others	1	0.21
Total			486	100.0

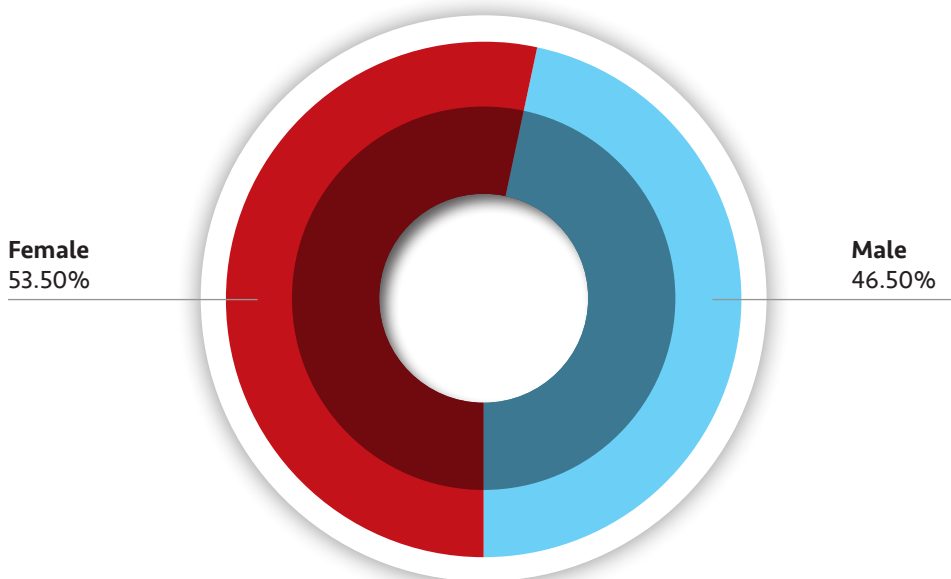
6.2.2 Gender

Majority of the patients in Kelantan are female, comprising 260 patients (53.50%). Meanwhile, 226 (46.50%) of the patients are male.

Table 6.6: Distribution of Patients in Kelantan by Gender

Centre	Male		Female	
	No.	%	No.	%
Hospital Raja Perempuan Zainab II, Kota Bharu	109	22.43	120	24.69
Hospital Universiti Sains Malaysia, Kubang Kerian	36	7.41	51	10.49
Hospital Pasir Mas	7	1.44	8	1.65
Hospital Gua Musang	12	2.47	8	1.65
Hospital Kuala Krai	20	4.12	29	5.97
Hospital Machang	4	0.82	13	2.67
Hospital Tanah Merah	19	3.91	20	4.12
Hospital Tengku Anis, Pasir Puteh	7	1.44	1	0.21
Hospital Jeli	6	1.23	3	0.62
Hospital Tumpat	6	1.23	7	1.44
Total	226	46.50	260	53.50

Figure 6.3: Distribution of Patients in Kelantan by Gender



6.2.3 Ethnic Group

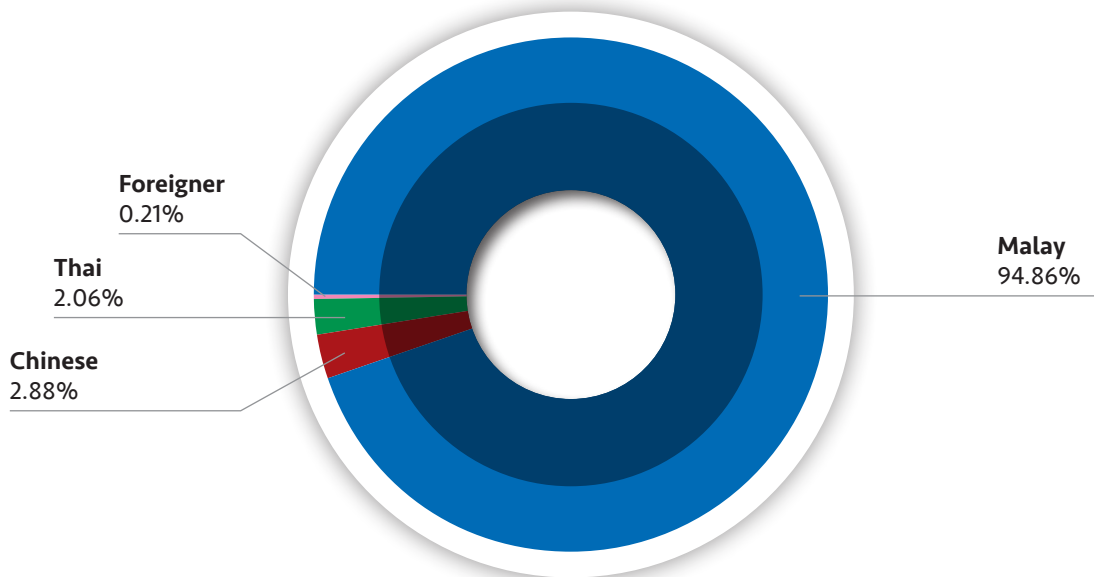
Table 6.7 shows the distribution of thalassaemia patients by ethnic group in Kelantan. Most thalassaemia patients in Kelantan are Malays (461 patients, 94.86%) followed by Chinese (14 patients, 2.88%), Thai (10 patients, 2.06%) and also one (0.21%) foreigner.

Table 6.7: Distribution of Patients in Kelantan by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	461	94.86
Chinese	14	2.88
Indian	0	0.00
Kadazan Dusun	0	0.00
Others:		
Thai	10	2.06
Foreigner	1	0.21
Total	486	100.0

6 Kelantan

Figure 6.4: Distribution of Patients in Kelantan by Ethnic Group



104

Table 6.8: Distribution of Patients in Kelantan According to Ethnic Group by Centre

Centre	Malay		Chinese		Thai		Foreigner	
	No.	%	No.	%	No.	%	No.	%
HRPZII, Kota Bharu	214	44.03	8	1.65	6	1.23	1	0.21
HUSM, Kubang Kerian	82	16.87	5	1.03	0	0.00	0	0.00
Hospital Kuala Krai	49	10.08	0	0.00	0	0.00	0	0.00
Hospital Machang	17	3.50	0	0.00	0	0.00	0	0.00
Hospital Pasir Mas	15	3.09	0	0.00	0	0.00	0	0.00
Hospital Gua Musang	19	3.91	1	0.21	0	0.00	0	0.00
Hospital Tanah Merah	39	8.02	0	0.00	0	0.00	0	0.00
Hospital Tengku Anis	8	1.65	0	0.00	0	0.00	0	0.00
Hospital Jeli	9	1.85	0	0.00	0	0.00	0	0.00
Hospital Tumpat	9	1.85	0	0.00	4	0.82	0	0.00
Total	461	94.86	14	2.88	10	2.06	1	0.21

6.3 Diagnosis

HbE-beta thalassaemia makes up the majority diagnosis of thalassaemia patients in Kelantan, with 241 patients (49.59%). This is followed by HbH disease (117 patients, 24.07%), beta thalassaemia major (70 patients, 14.40%) and beta thalassaemia intermedia (31 patients, 6.38%). The remaining 27 patients (5.56%) is of other diagnosis. Most of the patients have TDT.

Table 6.9: Distribution of Patients in Kelantan by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	70	14.40
Beta Thalassaemia Intermedia	31	6.38
HbE-Beta Thalassaemia	241	49.59
HbH Disease	117	24.07
Others*	27	5.56
Total	486	100.0

*Alpha thalassaemia, Hb Adana, delta-beta thalassaemia with concomitant alpha thalassaemia.

Figure 6.5: Distribution of Patients in Kelantan by Diagnosis

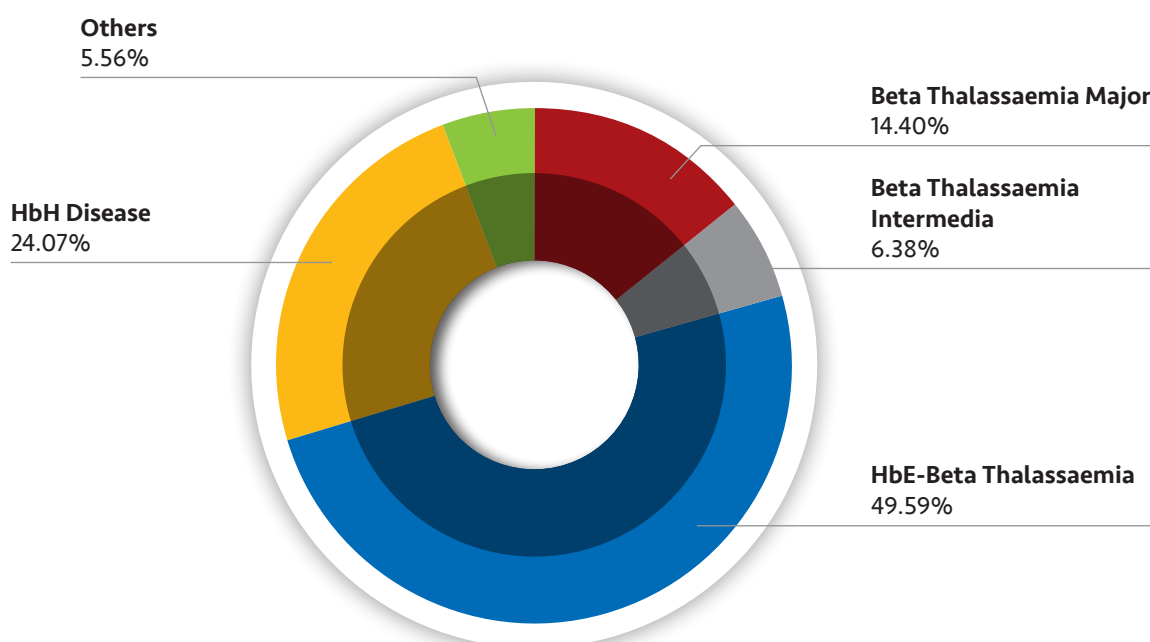


Table 6.10: Distribution of Patients in Kelantan According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
HRPZII, Kota Bharu	25	5.14	19	3.91	118	24.28	60	12.35	7	1.44
Hospital Pasir Mas	6	1.23	0	0.00	4	0.82	3	0.62	2	0.41
HUSM, Kubang Kerian	9	1.85	3	0.62	52	10.70	22	4.53	1	0.21
Hospital Kuala Krai	11	2.26	3	0.62	25	5.14	8	1.65	2	0.41
Hospital Machang	3	0.62	0	0.00	9	1.85	4	0.82	1	0.21
Hospital Gua Musang	7	1.44	0	0.00	9	1.85	3	0.62	1	0.21
Hospital Tanah Merah	4	0.82	4	0.82	11	2.26	11	2.26	9	1.85
Hospital Tengku Anis, Pasir Puteh	2	0.41	0	0.00	5	1.03	1	0.21	0	0.00
Hospital Jeli	3	0.62	0	0.00	4	0.82	1	0.21	1	0.21
Hospital Tumpat	0	0.00	2	0.41	4	0.82	4	0.82	3	0.62
Total	70	14.40	31	6.38	241	49.59	117	24.07	27	5.56

6 Kelantan

Based on Table 6.10, HRPZII has the highest number of thalassaemia patients in each type of diagnosis compared to other hospitals. This is due to HRPZII being a tertiary hospital in Kelantan. Thus, most of the thalassaemia patients in Kelantan prefer HRPZII for treatments.

Table 6.11: Distribution of Patients in Kelantan According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	70	Malay	66	13.58
		Chinese	2	0.41
		Thai	1	0.21
		Foreigner	1	0.21
Beta Thalassaemia Intermedia	31	Malay	30	6.17
		Chinese	1	0.21
		Thai	0	0.00
		Foreigner	0	0.00
HbE-Beta Thalassaemia	241	Malay	225	46.30
		Chinese	10	2.06
		Thai	6	1.23
		Foreigner	0	0.00
HbH Disease	117	Malay	114	23.45
		Chinese	0	0.00
		Thai	3	0.61
		Foreigner	0	0.00
Others	27	Malay	26	5.35
		Chinese	1	0.21
		Thai	0	0.00
		Foreigner	0	0.00
Total			486	100

Table 6.11 shows that Malay patients form the majority in all the four main types of thalassaemia (beta thalassaemia major, beta thalassaemia intermedia, HbE-beta thalassaemia and HbH disease) in Kelantan.

6.4 Treatment

6.4.1 Iron Chelation Therapy

Currently, there are three types of iron chelating agents available in the government hospitals: injection DFO (administered for a minimum of 8 hours via subcutaneous self-infusion), oral DFP and oral DFX.

Oral DFX is the newer agent among the three chelators. It was first made available in Malaysia in the late 2000s. Due to the high cost of the drug, its usage has been limited to paediatric patients or as second/third line drug to patients of older age groups, who developed side effects or fail to respond to the first line drugs (DFO or DFP).

Oral DFX is a dispersible tablet which can be taken once daily. Comparatively, it is easier to be taken by most patients.

Oral DFX became the most prescribed chelating agent (prescribed to 134 out of 276 patients, 48.55%) in Kelantan. The drug is mostly dispensed in hospitals with resident paediatricians.

Based on the Table 6.12, from a total 276 chelated patients, 134 patients (48.55%) are on oral DFX, 54 patients (19.57%) on oral DFP, 49 patients (17.75%) on subcutaneous DFO and the remaining 39 patients (14.12%) on combination therapy.

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

Iron Chelation	No. of Patients (n)	Percentage (%)
DFO only	49	17.75
DFP only	54	19.57
DFX only	134	48.55
DFO + DFP	18	6.52
DFP + DFX	6	2.17
DFO + DFX	14	5.07
DFO + DFP + DFX	1	0.36
Total	276	100.00

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

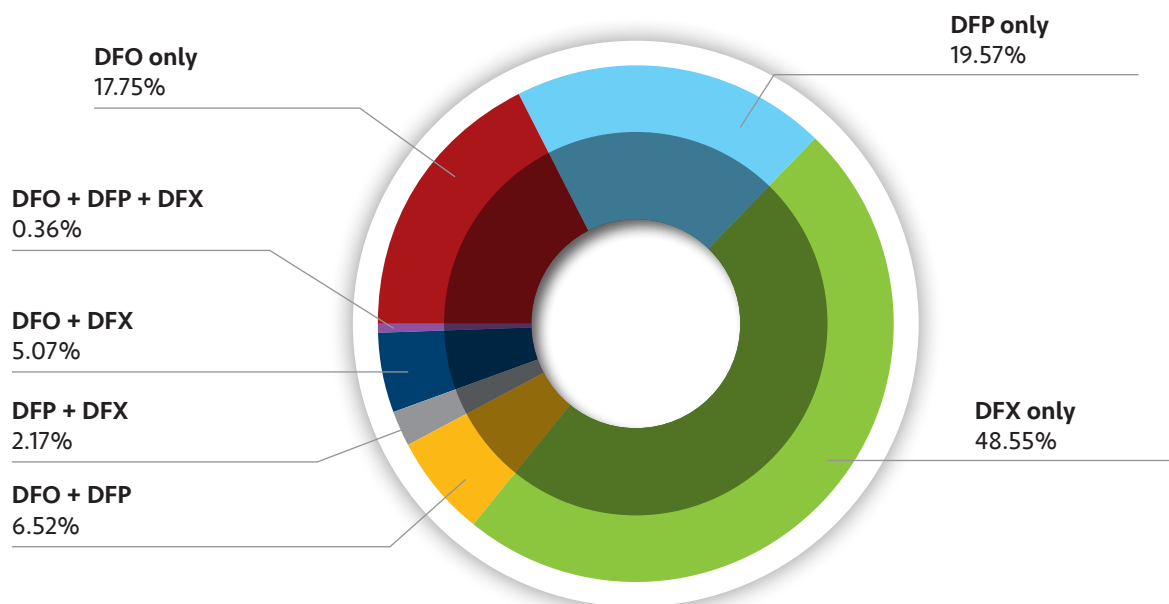
Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
HRPZII, Kota Bharu	131	DFO only	10	3.62
		DFP only	33	11.96
		DFX only	70	25.36
		DFO + DFP	7	2.54
		DFP + DFX	2	0.72
		DFO + DFX	8	2.90
		DFO + DFP + DFX	1	0.36
HUSM, Kubang Kerian	54	DFO only	21	7.61
		DFP only	11	3.99
		DFX only	6	2.17
		DFO + DFP	10	3.62
		DFP + DFX	1	0.36
		DFO + DFX	5	1.81
		DFO + DFP + DFX	0	0.00
Hospital Pasir Mas	6	DFO only	3	1.09
		DFP only	2	0.72
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	1	0.36
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

6 Kelantan

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Machang	13	DFO only	4	1.45
		DFP only	0	0.00
		DFX only	8	2.90
		DFO + DFP	0	0.00
		DFP + DFX	1	0.36
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kuala Krai	26	DFO only	6	2.17
		DFP only	2	0.72
		DFX only	16	5.80
		DFO + DFP	1	0.36
		DFP + DFX	1	0.36
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Gua Musang	16	DFO only	2	0.72
		DFP only	0	0.00
		DFX only	14	5.07
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tanah Merah	20	DFO only	0	0.00
		DFP only	5	1.81
		DFX only	15	5.43
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tengku Anis, Pasir Puteh	5	DFO only	3	1.09
		DFP only	0	0.00
		DFX only	2	0.72
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Tumpat	1	DFO only	0	0.00
		DFP only	1	0.36
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Jeli	4	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	3	1.09
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.36
		DFO + DFP + DFX	0	0.00
Total			276	100.00

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



6 Kelantan

Table 6.14: Distribution of Patients in Kelantan According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	6	DFO only	0	0.00
		DFP only	1	0.36
		DFX only	4	1.45
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.36
		DFO + DFP + DFX	0	0.00
5-9.9	41	DFO only	1	0.36
		DFP only	0	0.00
		DFX only	35	12.68
		DFO + DFP	0	0.00
		DFP + DFX	1	0.36
		DFO + DFX	4	1.45
		DFO + DFP + DFX	0	0.00
10-14.9	44	DFO only	7	2.54
		DFP only	3	1.09
		DFX only	31	11.23
		DFO + DFP	1	0.36
		DFP + DFX	0	0.00
		DFO + DFX	2	0.72
		DFO + DFP + DFX	0	0.00
15-19.9	60	DFO only	14	5.07
		DFP only	14	5.07
		DFX only	27	9.78
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	4	1.45
		DFO + DFP + DFX	1	0.36
20-24.9	48	DFO only	15	5.43
		DFP only	7	2.54
		DFX only	17	6.16
		DFO + DFP	5	1.81
		DFP + DFX	2	0.72
		DFO + DFX	2	0.72
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
25-29.9	35	DFO only	7	2.54
		DFP only	9	3.26
		DFX only	12	4.35
		DFO + DFP	6	2.17
		DFP + DFX	1	0.36
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30-34.9	14	DFO only	1	0.36
		DFP only	8	2.90
		DFX only	3	1.09
		DFO + DFP	1	0.36
		DFP + DFX	0	0.00
		DFO + DFX	1	0.36
		DFO + DFP + DFX	0	0.00
35-39.9	7	DFO only	1	0.36
		DFP only	2	0.72
		DFX only	1	0.36
		DFO + DFP	2	0.72
		DFP + DFX	1	0.36
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40-44.9	5	DFO only	1	0.36
		DFP only	3	1.09
		DFX only	1	0.36
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-49.9	10	DFO only	1	0.36
		DFP only	4	1.45
		DFX only	3	1.09
		DFO + DFP	2	0.72
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

6 Kelantan

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
50-54.9	3	DFO only	0	0.00
		DFP only	1	0.36
		DFX only	0	0.00
		DFO + DFP	1	0.36
		DFP + DFX	1	0.36
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60-64.9	1	DFO only	0	0.00
		DFP only	1	0.36
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	2	DFO only	1	0.36
		DFP only	1	0.36
		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			276	100.00

The youngest patient receiving iron chelation treatment is 3 years old and the eldest is 77 years old.

6.4.2 Serum Ferritin Level

There are 210 regularly transfused patients with their serum ferritin level measured and recorded in Kelantan. There are 43 patients (20.48%) with serum ferritin level < 1000 ng/mL, 81 patients (38.57%) with serum ferritin level 1000-2499 ng/mL, 52 patients (24.76%) with serum ferritin level 2500-4999 ng/mL, 26 patients (12.38%) with serum ferritin level 5000-9999 ng/mL, and 8 patients (3.81%) having serum ferritin level > 10000 ng/mL.

Table 6.15: Distribution of Patients in Kelantan by Most Recent Serum Ferritin Level

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
HUSM, Kubang Kerian	31	5	2.38	6	2.86	10	4.76	6	2.86	4	1.90
HRPZ II, Kota Bharu	123	30	14.29	53	25.24	24	11.43	13	6.19	3	1.43
Hospital Kuala Krai	12	3	1.43	5	2.38	2	0.95	1	0.48	1	0.48
Hospital Machang	8	1	0.48	3	1.43	1	0.48	3	1.43	0	0.00
Hospital Gua Musang	3	0	0.00	3	1.43	0	0.00	0	0.00	0	0.00
Hospital Tanah Merah	20	2	0.95	5	2.38	11	5.24	2	0.95	0	0.00
Hospital Tengku Anis, Pasir Puteh	2	0	0.00	1	0.48	1	0.48	0	0.00	0	0.00
Hospital Pasir Mas	3	0	0.00	0	0.00	3	1.43	0	0.00	0	0.00
Hospital Jeli	6	1	0.48	4	1.90	0	0.00	1	0.48	0	0.00
Hospital Tumpat	2	1	0.48	1	0.48	0	0.00	0	0.00	0	0.00
Total	210	43	20.48	81	38.57	52	24.76	26	12.38	8	3.81

6.5 Observation and Comments

- The current registry lacks an updated dataset on transfusion requirements, latest serum ferritin level, intake of iron chelators, complications, etc. This is due to few factors, such as:
 - No person-in-charge assigned specifically for handling thalassaemia patients at the district hospitals. Hospital staff do not understand the importance and needs of the MTR.
 - The staff do not implement good thalassaemia record management.
 - Poor record management; screening and test results were unorganised, and amount utilised for blood transfusions were not recorded (these issues occur mostly at medical units for adult patients in the district hospitals).
- The current data registry does not easily differentiate between TDTs and NDTTs. It also does not generate report on complications.

6.5.1 Recommendations

- A standardised chart of thalassaemia and patient diary can be introduced across the state and be made available for every thalassaemia patient. The chart is to be used in continuity. By implementing this step, data can be easily collected by the RA.
- Engagement of dedicated healthcare professionals in district hospitals to update the MTR.
- Further data cleaning is required.

6.5.2 Conclusions

- Total number of thalassaemia patients in Kelantan is 486 patients. Majority of thalassaemia patients received treatment at tertiary centres, i.e. HRPZII (229 patients out of 486, 47.12%) and HUSM (87 patients out of 486, 17.90%).
- In 2018, there were four deaths of thalassaemia patients recorded in the state.
- The female and male patient distribution slightly differs at 260 (53.50%) and 226 patients (46.50%), respectively.
- Most of the thalassaemia patients in Kelantan are Malays (461 patients, 94.86%).
- HbE-beta thalassaemia contributed the most number of patients (241 patients, 49.59%), followed by HbH disease (117 patients, 24.07%) and beta thalassaemia major (70 patients, 14.40%).
- DFX was the most prescribed chelating agent (134 patients, 48.55%).
- There are 210 regularly transfused patients who had their serum ferritin level measured in 2018. 124 patients (59.05%) have serum ferritin level < 2499 ng/mL and 86 patients (40.95%) have serum ferritin level > 2500 ng/mL.

7 Melaka

7.1 Introduction

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

Based on the Department of Statistics Malaysia, Melaka has a population of 922,200 as of 2018, comprising of Bumiputera (601,700, 65.25%), Chinese (including the Peranakan community; 216,900, 23.52%), Indians: (including the Chitty people; 51,100, 5.54%), other races (4,800, 0.52%), and non-Malaysian citizens (47,700, 5.17%).

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

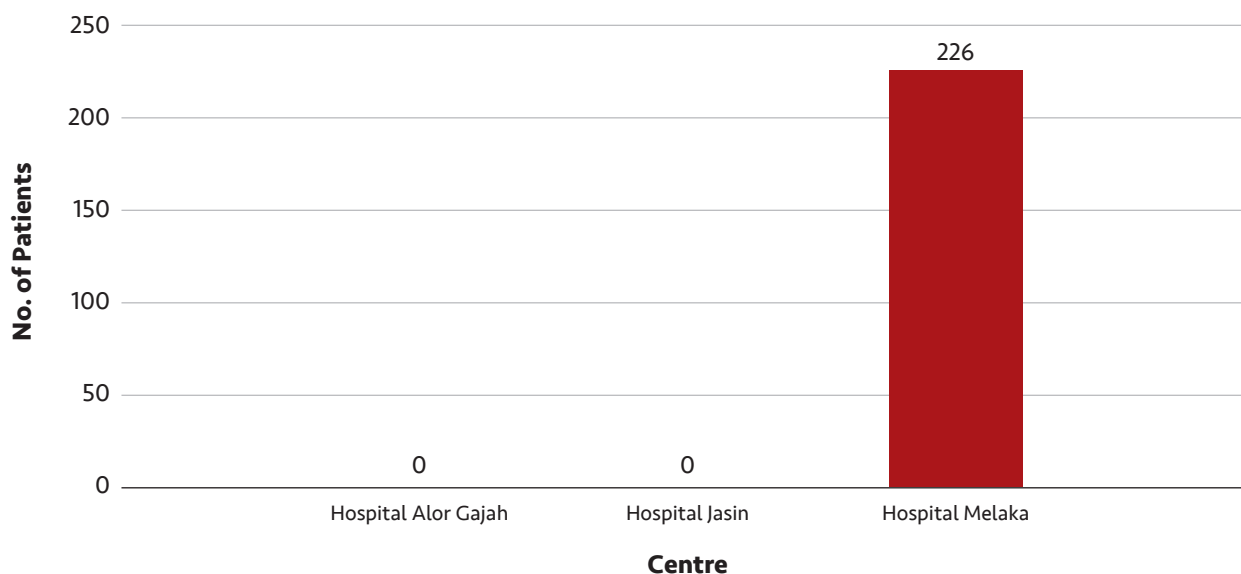
7.2 Patient Demographics

The total number of living patients is 226 in Melaka and the number of deceased patients up to the year 2018 is two. There was one patient death during admission to Hospital Melaka's intensive care unit in 2018.

Table 7.1: Distribution of Patients in Melaka by Centre

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
Hospital Alor Gajah	0	0.00
Hospital Jasin	0	0.00
Hospital Melaka	226	100.00
Total	226	100.00

Figure 7.1: Distribution of Patients in Melaka by Centre



**Table 7.2: Distribution of Patients in Melaka by Vital Status**

Vital Status	No. Of Patients
Alive	211
Cumulative Reported Cured by Stem Cell Therapy	10
Lost to Follow-up	5
Total	226
Death in 2018	1
Cumulative Reported Deaths	2

Table 7.3: Cumulative Causes of Death Since 2007 in Melaka

Cause of Death	No. of Patients
Endocrine complications	1
Others (ICU)	1
Total	2

7.2.1 Age

The youngest patient in Melaka is 1 year old and the eldest is 44 years old. The highest number of patients are in the age groups of 10-14.9 and 20-24.9 years old, both with 48 patients (21.24%) respectively. There are no patients above 45 years old in this state.

Table 7.4: Distribution of Patients in Melaka by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	26	11.50
5 - 9.9	45	19.91
10 -14.9	48	21.24
15 -19.9	29	12.83
20 - 24.9	48	21.24
25 - 29.9	22	9.73
30 - 34.9	7	3.10
35 - 39.9	0	0.00
40 - 44.9	1	0.44
45 - 49.9	0	0.00
50 - 54.9	0	0.00
55 - 59.9	0	0.00
60 - 64.9	0	0.00
Above 65	0	0.00
Total	226	100.00

7 Melaka

Figure 7.2: Distribution of Patients in Melaka by Age Group

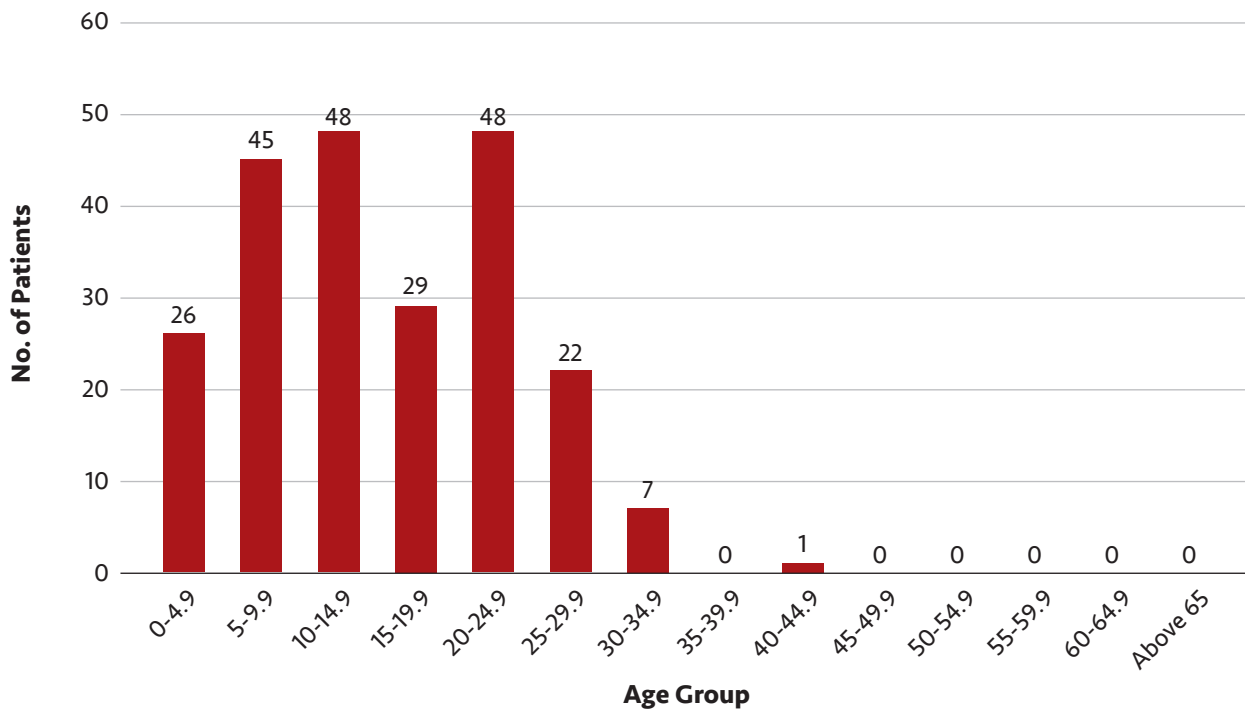


Table 7.5: Distribution of Patients in Melaka According to Diagnosis by Age Group

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	26	Beta Thalassaemia Major	7	3.10
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	10	4.42
		HbH Disease	7	3.10
		Others	2	0.88
5 - 9.9	45	Beta Thalassaemia Major	8	3.54
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	19	8.41
		HbH Disease	12	5.31
		Others	6	2.65
10 -14.9	48	Beta Thalassaemia Major	8	3.54
		Beta Thalassaemia Intermedia	3	1.33
		HbE-Beta Thalassaemia	17	7.52
		HbH Disease	15	6.64
		Others	5	2.21
15 -19.9	29	Beta Thalassaemia Major	6	2.65
		Beta Thalassaemia Intermedia	2	0.88
		HbE-Beta Thalassaemia	16	7.08
		HbH Disease	4	1.77
		Others	1	0.44

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
20 - 24.9	48	Beta Thalassaemia Major	16	7.08
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	21	9.29
		HbH Disease	10	4.42
		Others	1	0.44
25 - 29.9	22	Beta Thalassaemia Major	3	1.33
		Beta Thalassaemia Intermedia	1	0.44
		HbE-Beta Thalassaemia	12	5.31
		HbH Disease	5	2.21
		Others	1	0.44
30 - 34.9	7	Beta Thalassaemia Major	2	0.88
		Beta Thalassaemia Intermedia	1	0.44
		HbE-Beta Thalassaemia	3	1.33
		HbH Disease	1	0.44
		Others	0	0.00
35 - 39.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
40 - 44.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.44
		Others	0	0.00
Above 45	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			226	100.00

7 Melaka

7.2.2 Gender

Table 7.6 shows the distribution of thalassaemia patients in Melaka by gender. There are 116 (51.33%) male patients and 110 (48.67%) female patients in Melaka.

Figure 7.3: Distribution of Patients in Melaka by Gender

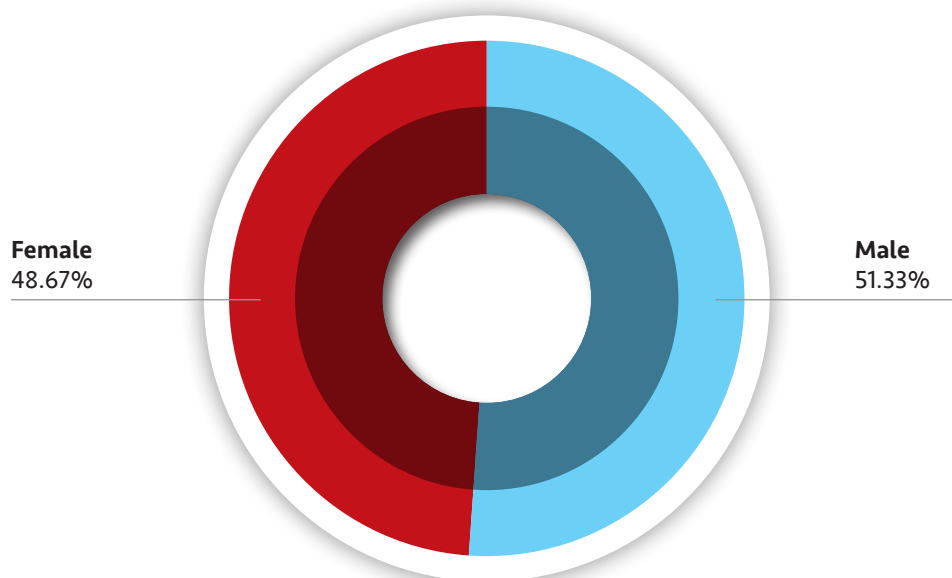


Table 7.6: Distribution of Patients in Melaka by Gender

Centre	Male		Female	
	No.	%	No.	%
Hospital Alor Gajah	0	0.00	0	0.00
Hospital Jasin	0	0.00	0	0.00
Hospital Melaka	116	51.33	110	48.67
Total	116	51.33	110	48.67

7.2.3 Ethnic Group

The ethnic group with the greatest number of patients in Melaka is Malay (178 patients, 78.76%), followed by Chinese (40 patients, 17.70%), Indian (3 patients, 1.33%), Bajau (1 patient, 0.44%), Iban (1 patient, 0.44%), Thai (2 patients, 0.88%) and Pribumi Sabah (1 patient, 0.44%).

Table 7.7: Distribution of Patients in Melaka by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	178	78.76
Chinese	40	17.70
Indian	3	1.33
Others:		
Bajau	1	0.44
Iban	1	0.44
Thai	2	0.88
Pribumi Sabah	1	0.44
Total	637	100

Figure 7.4: Distribution of Patients in Melaka by Ethnic Group

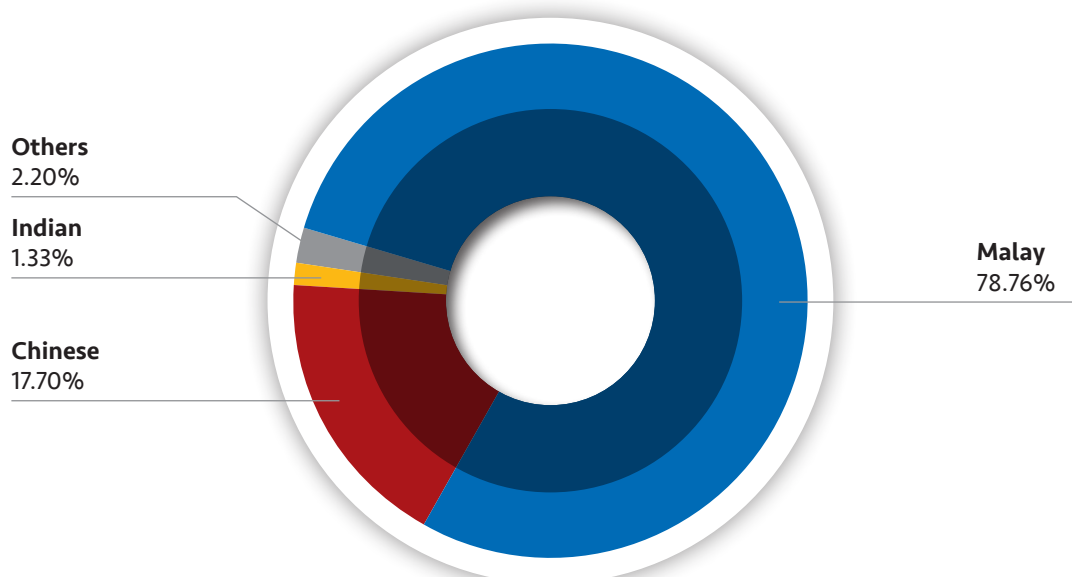


Table 7.8: Distribution of Patients in Melaka According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Bajau		Iban		Thai		Pribumi Sabah	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Alor Gajah	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Jasin	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Melaka	178	78.76	40	17.7	3	1.33	1	0.44	1	0.44	2	0.88	1	0.44
Total	178	78.76	40	17.7	3	1.33	1	0.44	1	0.44	2	0.88	1	0.44

7.3 Diagnosis

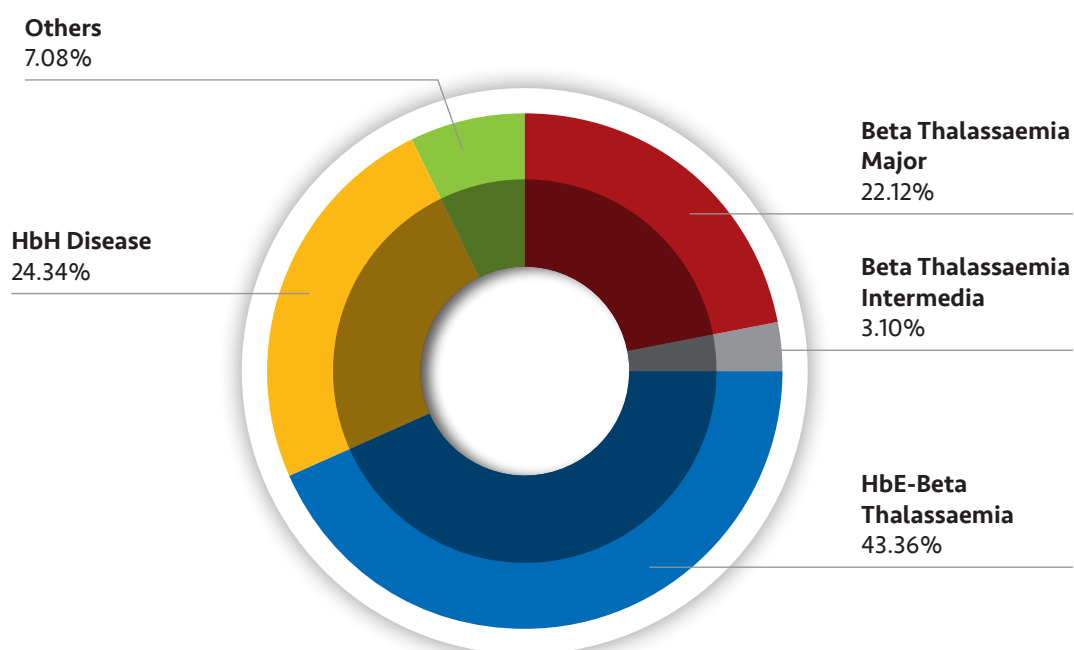
HbE-beta thalassaemia formed the majority diagnosis in Melaka with 98 patients. This is followed by HbH disease (55 patients), beta thalassaemia major (50 patients) and beta thalassaemia intermedia (7 patients).

Table 7.9: Distribution of Patients in Melaka by Diagnosis

Diagnosis	No. of Patients	Percentage (%)
Beta Thalassaemia Major	50	22.12
Beta Thalassaemia Intermedia	7	3.10
HbE-Beta Thalassaemia	98	43.36
HbH Disease	55	24.34
Others	16	7.08
Total	226	100.00

7 Melaka

Figure 7.5: Distribution of Patients in Melaka by Diagnosis



120

Table 7.10: Distribution of Patients in Melaka According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Alor Gajah	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Jasin	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Melaka	50	22.12	7	3.10	98	43.36	55	24.34	16	7.08
Total	50	22.12	7	3.10	98	43.36	55	24.34	16	7.08

Table 7.11: Distribution of Patients in Melaka According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	50	Malay	28	12.39
		Chinese	22	9.73
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Beta Thalassaemia Intermediate	7	Malay	6	2.65
		Chinese	0	0.00
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	1	0.44

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
HbE-Beta Thalassaemia	98	Malay	92	40.71
		Chinese	3	1.33
		Indian	2	0.88
		Kadazan-Dusun	0	0.00
		Others	1	0.44
HbH Disease	55	Malay	37	16.37
		Chinese	15	6.64
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	3	1.33
Others	16	Malay	15	6.64
		Chinese	0	0.00
		Indian	1	0.44
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Total			226	100.00

7.4 Treatment

7.4.1 Iron Chelation Therapy

The youngest patient in Melaka receiving iron chelation therapy is 3 years old (DFX). The total number of patients on iron chelators is 115 (50.88%). Out of this number, 50 patients (43.48%) receive DFX, 44 patients (38.26%) are on DFO, 16 patients (13.91%) receive a combination of iron chelators and 5 patients (4.35%) receive DFP.

Table 7.12: Distribution of Patients in Melaka by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	44	38.26
DFP only	5	4.35
DFX only	50	43.48
DFO + DFP	12	10.43
DFP + DFX	0	0.00
DFO + DFX	4	3.48
DFO + DFP + DFX	0	0.00
Total	115	100.00

Table 7.13: Distribution of Patients in Melaka According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Alor Gajah	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Jasin	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Melaka	115	DFO only	44	38.26
		DFP only	5	4.35
		DFX only	50	43.48
		DFO + DFP	12	10.43
		DFP + DFX	0	0.00
		DFO + DFX	4	3.48
		DFO + DFP + DFX	0	0.00
Total			115	100.00

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

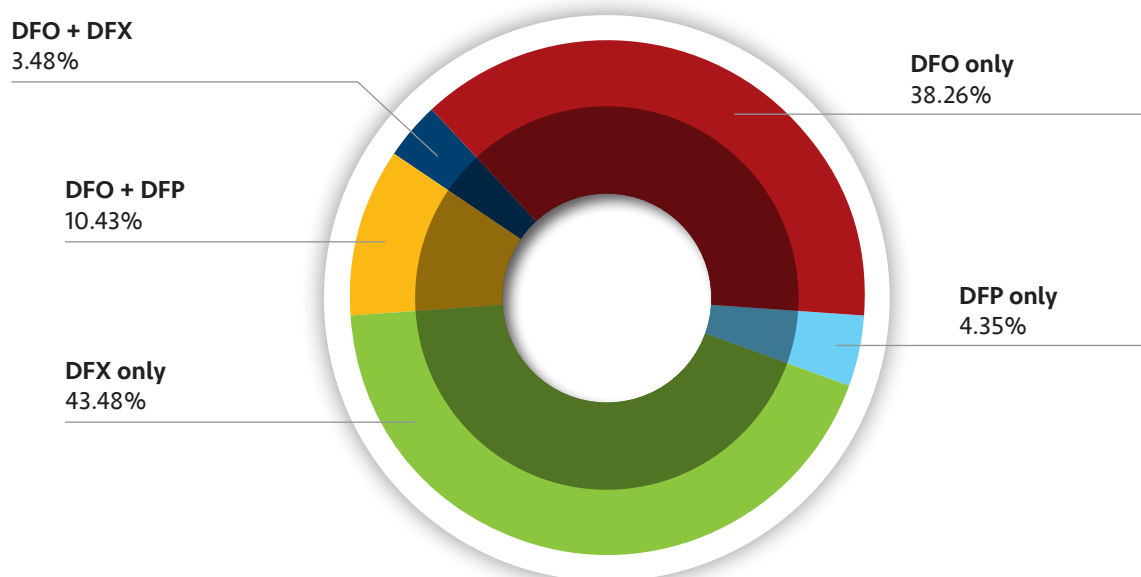


Table 7.14: Distribution of Patients in Melaka According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0 - 4.9	4	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	4	3.48
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	20	DFO only	4	3.48
		DFP only	0	0.00
		DFX only	13	11.30
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	3	2.61
		DFO + DFP + DFX	0	0.00
10 - 14.9	22	DFO only	4	3.48
		DFP only	0	0.00
		DFX only	18	15.65
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15 - 19.9	19	DFO only	7	6.09
		DFP only	1	0.87
		DFX only	9	7.83
		DFO + DFP	1	0.87
		DFP + DFX	0	0.00
		DFO + DFX	1	0.87
		DFO + DFP + DFX	0	0.00
20 - 24.9	33	DFO only	18	15.65
		DFP only	3	2.61
		DFX only	5	4.35
		DFO + DFP	7	6.09
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

7 Melaka

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
25 - 29.9	11	DFO only	6	5.22
		DFP only	1	0.87
		DFX only	0	0.00
		DFO + DFP	4	3.48
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30-34.9	6	DFO only	5	4.35
		DFP only	0	0.00
		DFX only	1	0.87
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
35-39.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40 - 44.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 45	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			115	100.00

7.4.2 Serum Ferritin Level

There are 60 patients in Melaka receiving regular transfusion who have their serum ferritin level measured. The lowest serum ferritin level is 219.3 ng/mL (2-year-old patient) and the highest is 11,333.00 ng/mL (16-year-old patient). The percentage of patients with serum ferritin level lower than 2499 ng/mL is 61.67%, whereas percentage of patients with serum ferritin level above 2500 ng/mL is 38.33%.

Table 7.15: Distribution of Patients in Melaka by Most Recent Serum Ferritin Level

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Alor Gajah	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Jasin	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Melaka	60	13	21.67	24	40.00	17	28.33	5	8.33	1	1.67
Total	60	13	21.67	24	40.00	17	28.33	5	8.33	1	1.67

7.5 Observation and Comments

The number of living patients currently registered at Hospital Melaka is 226. Of these, 211 are active patients, 10 patients underwent BMT and 5 patients is lost to follow-up. No new thalassaemia births were recorded in 2018, whereas five patients were born in 2017.

Majority of patients in Melaka are in the age group of 10-14.9 or 20-24.9 years old. The patients' age ranges from 1 year old to 44 years old. Out of the 226 thalassaemia patients in Melaka, 119 patients (52.65%) are under 15 years old and falls under paediatric category. Most of the patients are male, with the female to male ratio of 1:1.05. Malays constitute 78.76% of the total number of patients, followed by Chinese (17.70%), Indian (1.33%), Thai (0.88%), Bajau, Iban and Pribumi Sabah (each at 0.44%).

Most of the TDT patients in Melaka were diagnosed with HbE-beta thalassaemia (43.36%), followed by beta thalassaemia major (22.12%). On the other hand, the highest number of NTDT patients were diagnosed HbH disease (including HbH Constant Spring; 24.34%), followed by beta thalassaemia intermedia (3.10%). Other diagnoses such as alpha thalassaemia, delta beta thalassaemia and HbS with beta thalassaemia forms 7.08% of the total patient number in Melaka. The oldest surviving beta thalassaemia major patient is in the age range of 30-34.9 years old.

Despite chelation and monitoring, only 30% of the patients have their serum ferritin level measured. Only 61.67% of this group of patients have a serum ferritin level below 2499 ng/mL. Overall, there is still a need to continue to educate, encourage and provide information to our patients to improve their iron status and their overall clinical outcome.

8 Negeri Sembilan

8.1 Introduction

Negeri Sembilan, is one of the 14 states which constitutes Malaysia. It is located on the western coast of Peninsular Malaysia, just south of Kuala Lumpur and is bordered in the north by Selangor, in the east by Pahang and in the south by Melaka and Johor.

Negeri Sembilan has collective population of 1.13 million in 2018 and consists of seven districts, namely Seremban, Port Dickson, Kuala Pilah, Jempol, Jelebu, Rembau and Tampin.

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

The total number of thalassaemia patients registered in the MTR in Negeri Sembilan are 181 patients. Most patients (135 patients) received treatment at HTJ, followed by HTAN (37 patients). HTJ is the only hospital in Negeri Sembilan which provides day care service including blood transfusion for thalassaemia patients.

8.2 Patient Demographics

Patient were categorised as alive, lost to follow-up and cured by transplant. The total number of living patients is 181 and the number of deceased patient up to 2018 is 6 patients.

Table 8.1: Distribution of Patients in Negeri Sembilan by Centre

Centre	No. of Patients (n)	Percentage (%)
Hospital Jelebu	6	3.31
Hospital Tuanku Ampuan Najihah, Kuala Pilah	37	20.44
Hospital Port Dickson	3	1.66
Hospital Tuanku Ja'afar, Seremban	135	74.59
Hospital Tampin	0	0.00
Hospital Jempol	0	0.00
Total	181	100.00



Figure 8.1: Distribution of Patients in Negeri Sembilan by Centre

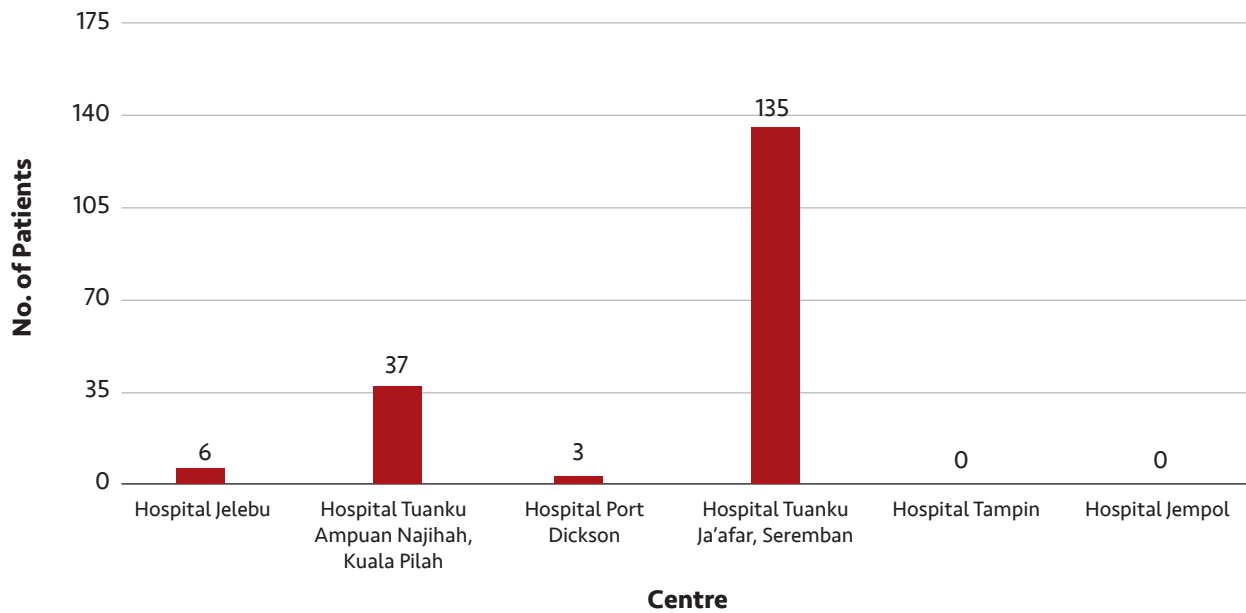


Table 8.2: Distribution of Patients in Negeri Sembilan by Vital Status

Vital Status	No. Of Patients
Alive	172
Cumulative Reported Cured by Stem Cell Therapy	9
Lost to Follow-up	0
Total	181
Deaths in 2018	0
Cumulative Reported Deaths	6

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

Cause of Death	No. of Patients
Infections	1
Cardiac Causes	2
Liver Disease	1
Endocrine Complications	1
Unknown*	1
Total	6

*Missing data.

8.2.1 Age

The youngest patient in Negeri Sembilan is a 2-year-old and the oldest is 63 years old. The age group with the highest number of patients is the 15-19.9 years old category.

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

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	9	4.97
5 - 9.9	30	16.57
10 -14.9	18	9.94
15 -19.9	37	20.44
20 - 24.9	27	14.92
25 - 29.9	19	10.50
30 - 34.9	13	7.18
35 - 39.9	7	3.87
40 - 44.9	7	3.87
45 - 49.9	5	2.76
50 - 54.9	3	1.66
55 - 59.9	3	1.66
60 - 64.9	3	1.66
Above 65	0	0.00
Total	181	100.00

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

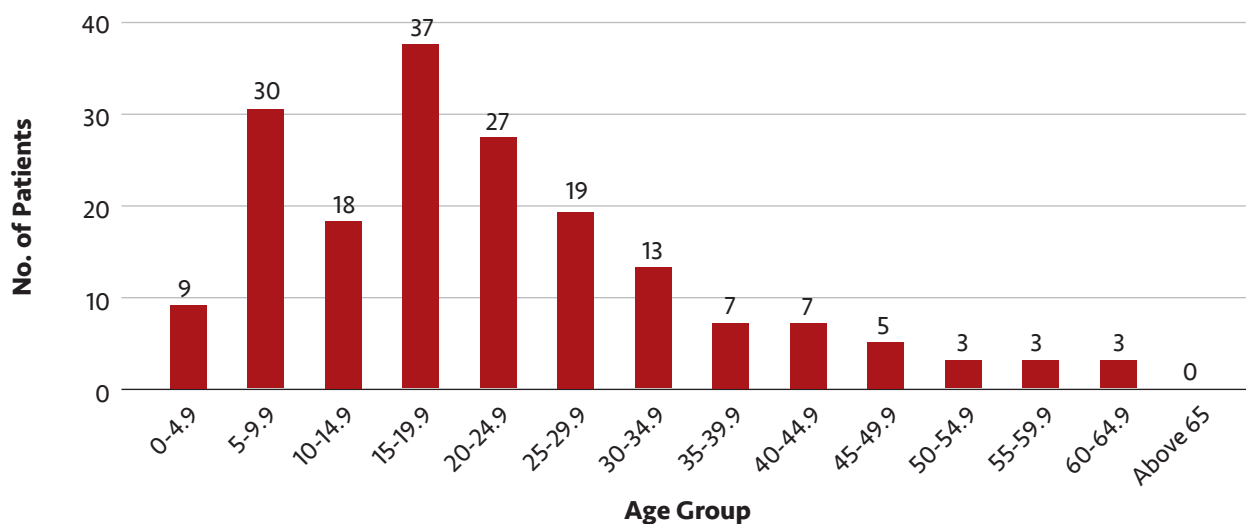


Table 8.5: Distribution of Patients in Negeri Sembilan According to Diagnosis by Age Group

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	9	Beta Thalassaemia Major	3	1.66
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	5	2.76
		HbH Disease	1	0.55
		Others*	0	0.00
5 - 9.9	30	Beta Thalassaemia Major	9	4.97
		Beta Thalassaemia Intermedia	4	2.21
		HbE-Beta Thalassaemia	9	4.97
		HbH Disease	4	2.21
		Others*	4	2.21
10 - 14.9	18	Beta Thalassaemia Major	4	2.21
		Beta Thalassaemia Intermedia	1	0.55
		HbE-Beta Thalassaemia	9	4.97
		HbH Disease	3	1.66
		Others*	1	0.55
15 - 19.9	37	Beta Thalassaemia Major	11	6.08
		Beta Thalassaemia Intermedia	2	1.10
		HbE-Beta Thalassaemia	13	7.18
		HbH Disease	6	3.31
		Others*	5	2.76
20 - 24.9	27	Beta Thalassaemia Major	8	4.42
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	15	8.29
		HbH Disease	3	1.66
		Others*	1	0.55
25 - 29.9	19	Beta Thalassaemia Major	6	3.31
		Beta Thalassaemia Intermedia	1	0.55
		HbE-Beta Thalassaemia	6	3.31
		HbH Disease	4	2.21
		Others*	2	1.10
30 - 34.9	13	Beta Thalassaemia Major	7	3.87
		Beta Thalassaemia Intermedia	2	1.10
		HbE-Beta Thalassaemia	2	1.10
		HbH Disease	1	0.55
		Others*	1	0.55

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	7	Beta Thalassaemia Major	1	0.55
		Beta Thalassaemia Intermedia	3	1.66
		HbE-Beta Thalassaemia	2	1.10
		HbH Disease	0	0.00
		Others*	1	0.55
40 - 44.9	7	Beta Thalassaemia Major	1	0.55
		Beta Thalassaemia Intermedia	2	1.10
		HbE-Beta Thalassaemia	3	1.66
		HbH Disease	1	0.55
		Others*	0	0.00
45 - 49.9	5	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	3	1.66
		HbE-Beta Thalassaemia	1	0.55
		HbH Disease	1	0.55
		Others*	0	0.00
50 - 54.9	3	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	1.10
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.55
		Others*	0	0.00
55 - 59.9	3	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.55
		HbE-Beta Thalassaemia	1	0.55
		HbH Disease	1	0.55
		Others*	0	0.00
60 - 64.9	3	Beta Thalassaemia Major	1	0.55
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	1	0.55
		HbH Disease	1	0.55
		Others*	0	0.00
Above 65	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others*	0	0.00
Total			181	100.00

*Alpha thalassaemia, HbH Constant Spring, pure red cell aplasia and sickle cell disease heterozygous.

8.2.2 Gender

Figure 8.3 shows the distribution of patients in Negeri Sembilan by gender. 85 patients (46.96%) in Negeri Sembilan are male and 96 patients (53.04%) are female.

Figure 8.3: Distribution of Patients in Negeri Sembilan by Gender

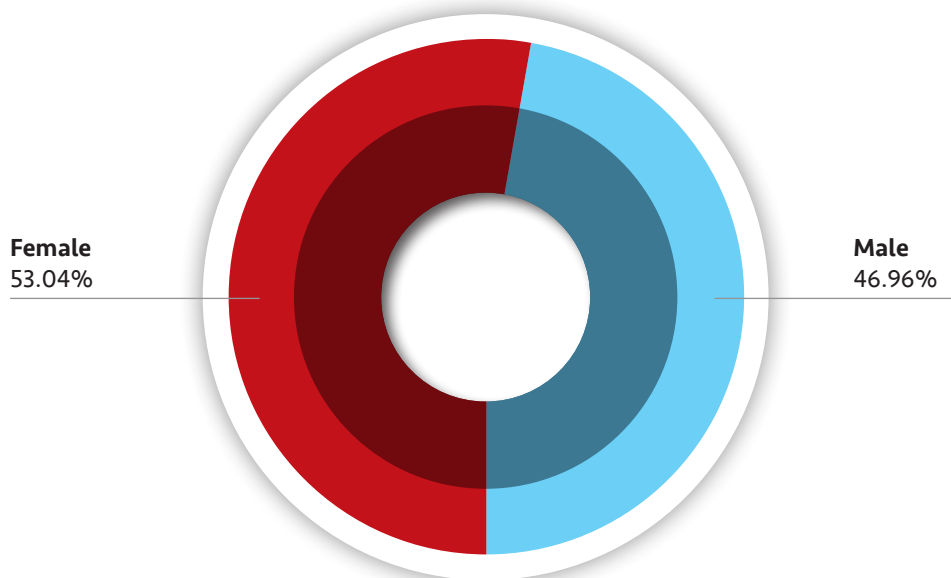


Table 8.6: Distribution of Patients in Negeri Sembilan According to Gender by Centre

Centre	Male		Female	
	No.	%	No.	%
Hospital Jelebu	3	1.66	3	1.66
Hospital Tuanku Ampuan Najihah, Kuala Pilah	26	14.36	11	6.08
Hospital Port Dickson	1	0.55	2	1.10
Hospital Tuanku Ja'afar	55	30.39	80	44.20
Hospital Tampin	0	0.00	0	0.00
Hospital Jempol	0	0.00	0	0.00
Total	85	46.96	96	53.04

8.2.3 Ethnic Group

The distribution of thalassaemia patients in Negeri Sembilan according to ethnic group are as follows: 141 out of 181 total number of patients (78.00%) are Malay, 29 patients (16.00%) are Chinese, 3 patients (1.70%) are Indian, 1 patient (0.60%) is Kadazan-Dusun, 6 patients (3.30%) are Orang Asli and 1 Thai patient (0.60%).

Table 8.7: Distribution of Patients in Negeri Sembilan by Ethnic Group

Ethnic Group	No of Patients (n)	Percentage (%)
Malay	141	77.90
Chinese	29	16.02
Indian	3	1.66
Kadazan-Dusun	1	0.55
Orang Asli	6	3.31
Others:		
Thai	1	0.55
Total	181	100.00

Figure 8.4: Distribution of Patients in Negeri Sembilan by Ethnic Group

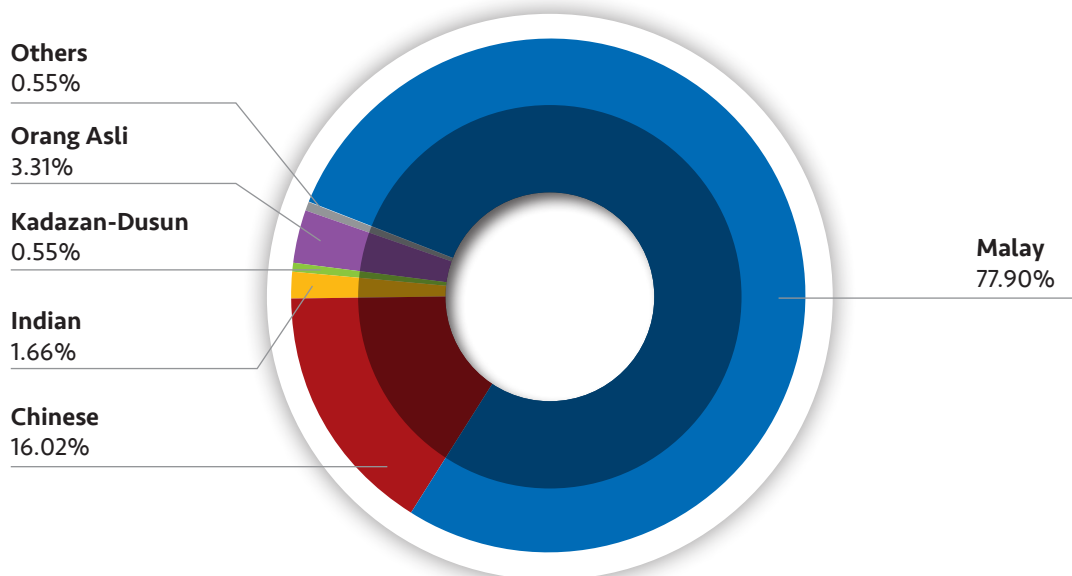


Table 8.8: Distribution of Patients in Negeri Sembilan According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Kadazan-Dusun		Orang Asli		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Jelebu	6	3.31	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Tuanku Ampuan Najihah	26	14.36	4	2.21	1	0.55	0	0.00	6	3.31	0	0.00
Hospital Port Dickson	2	1.10	0	0.00	0	0.00	0	0.00	0	0.00	1	0.55
Hospital Tuanku Ja'afar	107	59.12	25	13.81	2	1.10	1	0.55	0	0.00	0	0.00
Hospital Tampin	0	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	141	77.90	29	16.02	3	1.66	1	0.55	6	3.31	1	0.55

8.3 Diagnosis

HbE-beta thalassaemia formed the majority diagnosis with 67 patients. This is followed by beta thalassaemia major (51 patients), HbH disease (27 patients) and beta thalassaemia intermedia (21 patients).

Table 8.9: Distribution of Patients in Negeri Sembilan by Diagnosis

Diagnosis	No. of Patients	Percentage (%)
Beta Thalassaemia Major	51	28.18
Beta Thalassaemia Intermedia	21	11.60
HbE-Beta Thalassaemia	67	37.02
HbH Disease	27	14.92
Others	15	8.29
Total	181	100.00

Figure 8.5: Distribution of Patients in Negeri Sembilan by Diagnosis

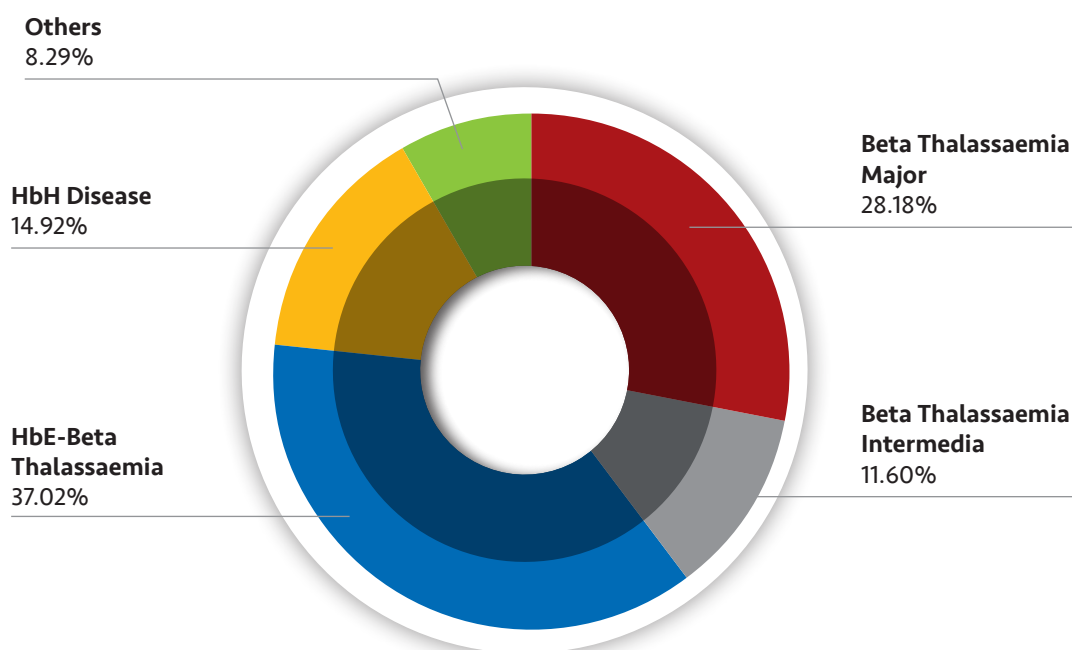


Table 8.10: Distribution of Patients in Negeri Sembilan According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Jelebu	4	2.21	0	0.00	2	1.10	0	0.00	0	0.00
Hospital Tuanku Ampuan Najihah	5	2.76	0	0.00	13	7.18	8	4.42	11	6.08
Hospital Port Dickson	0	0.00	0	0.00	3	1.66	0	0.00	0	0.00
Hospital Tuanku Ja'afar	42	23.20	21	11.60	49	27.07	19	10.50	4	2.21
Hospital Tampin	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	51	28.18	21	11.60	67	37.02	27	14.92	15	8.29

8 Negeri Sembilan

Table 8.11: Distribution of Patients in Negeri Sembilan According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	51	Malay	34	18.78
		Chinese	16	8.84
		Indian	0	0.00
		Kadazan-Dusun	1	0.55
		Others	0	0.00
Beta Thalassaemia Major	21	Malay	17	9.39
		Chinese	4	2.21
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
HbE-Beta Thalassaemia	67	Malay	61	33.70
		Chinese	3	1.66
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	3	1.66
HbH Disease	27	Malay	20	11.05
		Chinese	3	1.66
		Indian	2	1.10
		Kadazan-Dusun	0	0.00
		Others	2	1.10
Others	15	Malay	9	4.97
		Chinese	3	1.66
		Indian	1	0.55
		Kadazan-Dusun	0	0.00
		Others	2	1.10
Total			181	100.00

8.4 Treatment

8.4.1 Iron Chelation Therapy

Out of 181 patients in Negeri Sembilan, 115 patients (63.54%) are receiving regular transfusion. They are mainly patients with thalassaemia major or HbE-beta thalassaemia. Most patients receive DFO (33 patients, 28.70%), followed by DFX (31 patients, 26.96%), DFP (27 patients, 23.48%) and a combination therapy (24 patients, 20.87%).

Table 8.12: Distribution of Patients in Negeri Sembilan by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	33	28.70
DFP only	27	23.48
DFX only	31	26.96
DFO + DFP	20	17.39
DFP + DFX	0	0.00
DFO + DFX	4	3.48
DFO + DFP + DFX	0	0.00
Total	115	100.00

Table 8.13: Distribution of Patients in Negeri Sembilan According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Jelebu	5	DFO only	4	3.48
		DFP only	1	0.87
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Tuanku Ampuan Najihah, Kuala Pilah	18	DFO only	4	3.48
		DFP only	4	3.48
		DFX only	9	7.83
		DFO + DFP	1	0.87
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Port Dickson	3	DFO only	1	0.87
		DFP only	0	0.00
		DFX only	2	1.74
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

8 Negeri Sembilan

136

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Tuanku Ja'afar	89	DFO only	24	20.87
		DFP only	22	19.13
		DFX only	20	17.39
		DFO + DFP	19	16.52
		DFP + DFX	0	0.00
		DFO + DFX	4	3.48
		DFO + DFP + DFX	0	0.00
Hospital Tampin	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Jempol	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			115	100.00

Figure 8.6: Distribution of Patients in Negeri Sembilan by Type of Iron Chelator Received

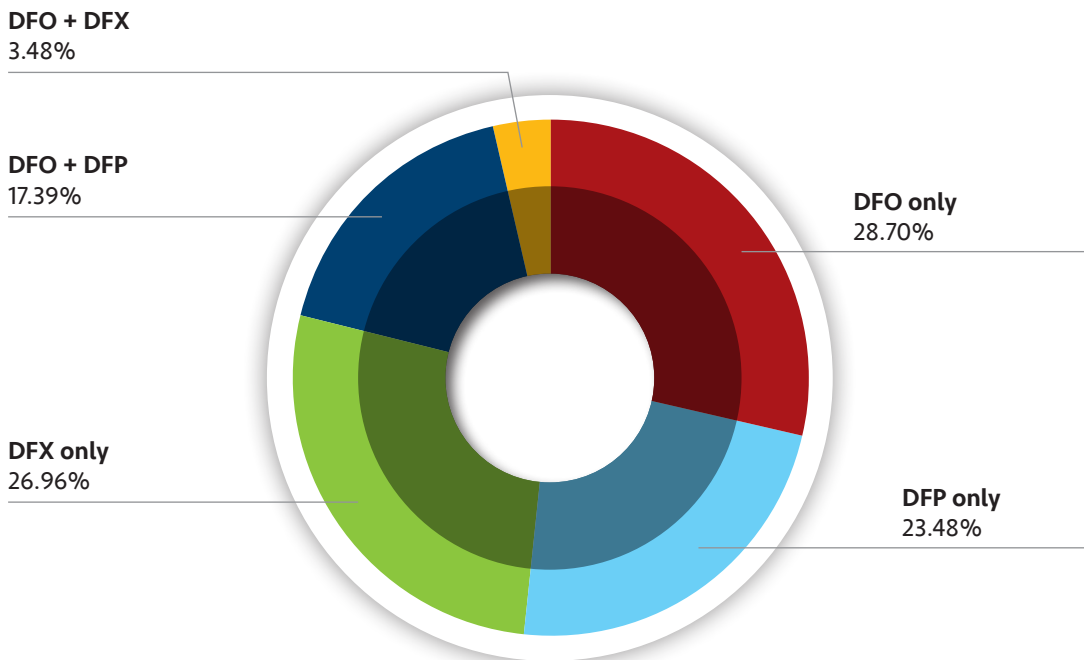


Table 8.14: Distribution of Patients in Negeri Sembilan According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0 - 4.9	4	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	4	3.48
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
5 - 9.9	22	DFO only	3	2.61
		DFP only	1	0.87
		DFX only	17	14.78
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.87
10 - 14.9	8	DFO only	1	0.87
		DFP only	1	0.87
		DFX only	5	4.35
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.87
15 - 19.9	26	DFO only	12	10.43
		DFP only	1	0.87
		DFX only	3	2.61
		DFO + DFP	8	6.96
		DFP + DFX	0	0.00
		DFO + DFX	2	1.74
20 - 24.9	22	DFO only	8	6.96
		DFP only	6	5.22
		DFX only	1	0.87
		DFO + DFP	7	6.09
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
25 - 29.9	10	DFO only	3	2.61
		DFP only	5	4.35
		DFX only	0	0.00
		DFO + DFP	2	1.74
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00

8 Negeri Sembilan

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
30-34.9	10	DFO only	2	1.74
		DFP only	6	5.22
		DFX only	1	0.87
		DFO + DFP	1	0.87
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
35-39.9	3	DFO only	2	1.74
		DFP only	1	0.87
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
40 - 44.9	3	DFO only	1	0.87
		DFP only	1	0.87
		DFX only	0	0.00
		DFO + DFP	1	0.87
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
45 - 49.9	4	DFO only	0	0.00
		DFP only	4	3.48
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
50 - 54.9	1	DFO only	0	0.00
		DFP only	1	0.87
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
55 - 59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
60-64.9	2	DFO only	1	0.87
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	0.87
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
Above 65	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
Total			115	100.00

8.4.2 Serum Ferritin Level

Table 8.15 shows that 61 regularly transfused patients (33.70%) in Negeri Sembilan have their serum ferritin measured. The lowest ferritin level recorded is 226.31 ng/mL (35-year-old patient) and the highest is 36,496.75 ng/mL (26-year-old patient). The percentage of patients with serum ferritin level lower than 2499 ng/mL is 39.34%, whereas the percentage of patients with serum ferritin above 2500 ng/mL are 60.66%.

Table 8.15: Distribution of Patients in Negeri Sembilan by Most Recent Serum Ferritin Level

Centre	Serum Ferritin Level (ng/mL)	< 1000		1000-2499		2500-4999		5000-9999		10,000+		
		Total	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Jelebu	0	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Tuanku Ampuan Najihah, Kuala Pilah	6	0	0	0.00	1	1.64	3	4.92	1	1.64	1	1.64
Hospital Port Dickson	2	0	0	0.00	2	3.28	0	0.00	0	0.00	0	0.00
Hospital Tuanku Jaafar	53	8	8	13.11	13	21.31	20	32.79	7	11.48	5	8.20
Hospital Tampin	0	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	61	8	8	13.11	16	26.23	23	37.70	8	13.11	6	9.84

8.5. Observation and Comments

The current registry in Negeri Sembilan lacks up-dated information on transfusion requirement, most recent serum ferritin level, intake of iron chelator and complications. These are due to a few factors such as:

- ✓ No person-in-charge assigned specifically for handling thalassaemia patients at the district hospitals. Hospital staff do not understand the importance and needs of MTRs.
- ✓ The staff do not implement good thalassaemia management.
- ✓ Poor record management; screening and test results were unorganised, and amount utilised for blood transfusions were not recorded (these issues occur mostly at medical units for adult patients, i.e. HT) and district hospitals).

In the process of generating the annual report, we detected a certain number of patients who were diagnosed as alpha thalassaemia were actually alpha trait (based on DNA analysis result). The current data registry does not differentiate between TDTs and NTDTs. It also does not generate report on complications.

8.5.1 Conclusion

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

The age group with the highest number of thalassaemia patients in Negeri Sembilan is between 15-29.9 years old. The patients' age ranges between 2 to 61 years old. 57 patients (31.49%) are below 15 years of age and is under paediatric care. Majority of the patients in Negeri Sembilan are females, with the male to female ratio of 1:1.13. Malays constitute 77.90% of the total number of patients, followed by Chinese (16.02%), Orang Asli (3.31%), Indian (1.66%), Kadazan-Dusun (0.55%) and other ethnicities (0.55%).

TDT patients in Negeri Sembilan are typically diagnosed with HbE-beta thalassaemia (37.02%) followed by beta thalassaemia major (28.18%). On the other hand, NTDT patients were diagnosed with HbH disease (14.92%), followed by beta thalassaemia intermedia (11.60%). Other diagnoses, such as alpha thalassaemia, HbH Constant Spring, pure red cell aplasia and sickle cell disease heterozygous forms 8.29% of the diagnoses. The oldest surviving beta thalassaemia major patient is in the range of 60-64.9 years old.

Only 63.54% out of the 181 patients in Negeri Sembilan are receiving iron chelation therapy, out of which 29.57% are under paediatric care. The remaining 36.46% are not receiving any form of chelator.

Finally, despite chelation and monitoring, only 33.70% of the patients have their serum ferritin level measured. Of that amount, only 13.11% of patients have serum ferritin level below 1000 ng/mL, and 26.23% of patients have a serum ferritin level between 1000-2499 ng/mL.

9 Pahang

9.1. Introduction

The state of Pahang has a total of 11 hospitals; these consist of a state hospital (Hospital Tengku Ampuan Afzan, [HTAA], Kuantan), 1 major hospital with specialist (Hospital Sultan Haji Ahmad Shah [HOSHAS], Temerloh), 2 minor hospitals with specialist (Hospital Kuala Lipis and Hospital Pekan) and 7 district hospitals.

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

The total population in Pahang according to the Department of Statistics Malaysia is 1.67 million, as of January 2018. There are a total of 437 thalassaemia patients in the state of Pahang.

Transfusion facilities has markedly improved in HTAA and HOSHAS, where all patients may receive blood transfusion in day care units and receive filtered and NAT-tested blood. Other district hospitals still require ward admissions for transfusion and also receive filtered blood.

With the latest advances in thalassaemia service, HTAA is now able to provide MRI T2* facility since October 2017. Bone densitometry is also provided under the radiology department in HTAA.

9.2. Patient Demographics

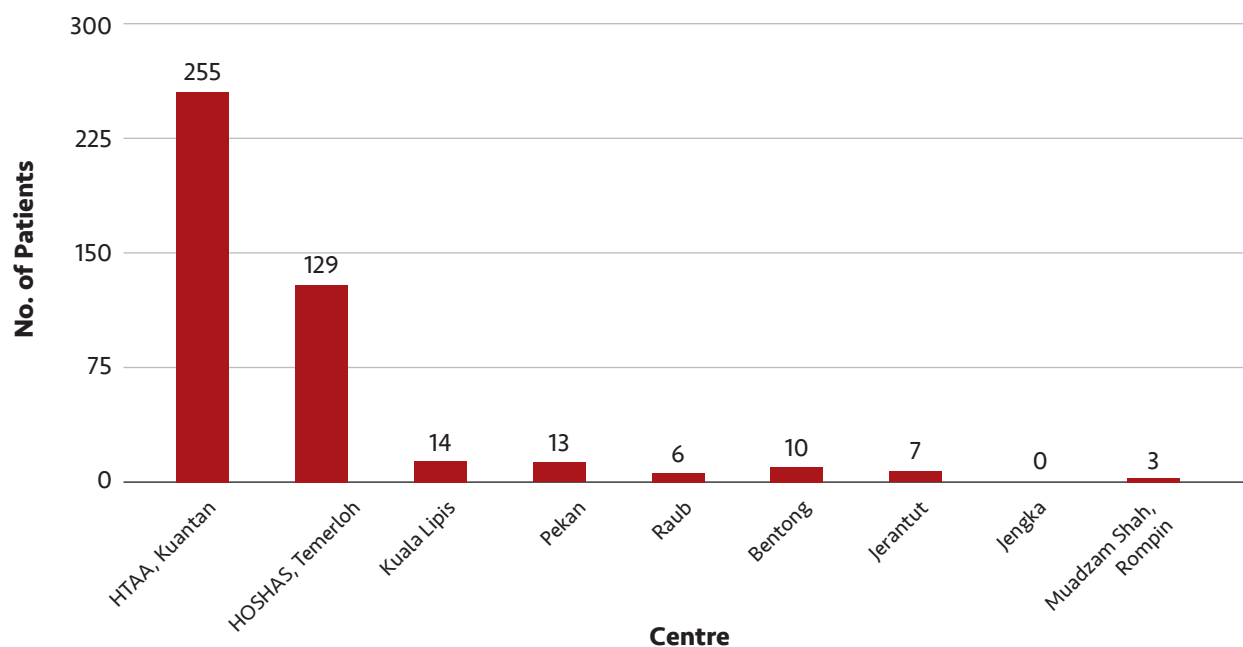
Data analysed were taken from patients with status of either alive, lost to follow-up or cured by transplant, and excludes deceased patients. Total number of living patients in Pahang are 437 patients, only one patient was cured by transplant and 3 deaths were reported in 2018. Two of the three deaths were due to infection and one due to cardiac arrhythmia.

Table 9.1: Distribution of Patients in Pahang by Centre

Centre	Patients Distribution	
	No. of Patients (n)	Percentage (%)
Tengku Ampuan Afzan, Kuantan (Hospital with specialist)	255	58.35
Sultan Haji Ahmad Shah, Temerloh (Hospital with specialist)	129	29.52
Kuala Lipis (Hospital with specialist)	14	3.20
Pekan (Hospital with specialist)	13	2.97
Raub (Covered by HOSHAS)	6	1.37
Bentong (Covered by HOSHAS)	10	2.29
Jerantut (Covered by HOSHAS)	7	1.60
Jengka (Covered by HOSHAS)	0	0.00
Muadzam Shah, Rompin (Covered by HTAA / Pekan)	3	0.69
Total	437	100.00

9 Pahang

Figure 9.1: Distribution of Patients in Pahang by Centre



Majority of thalassaemia patients received their treatment at tertiary centres, i.e. HTAA (255 out of 437 patients (58.3%) and HOSHAS (129 out of 437 patients (29.52%).

Table 9.2: Distribution of Patients in Pahang by Vital Status

Vital Status	No. of Patients
Alive	399
Cumulative Reported Cured by Stem Cell Therapy	1
Lost to Follow-up	37
Total	437
Deaths in 2018	3
Cumulative Reported Deaths	36

One of the three deaths in 2018 was of a 24-year-old Malay woman (HTAA patient) due to cardiac arrhythmia. The two other deaths were of a 29- and a 64-year-old Malay women (HOSHAS patients), due to sepsis.

Table 9.3: Cumulative Causes of Death Since 2007 in Pahang

Causes of Death	No. of Patients
Cardiac causes	6
Infections (2 cases died during BMT)	15
Arrhythmias	2
Liver disease	1
Endocrine complications	0
Thrombosis	0
Tumours	0
Cholecystectomy	0
Others:	
Accident	1
Fall in toilet	1
HIV	1
Advanced malignancy of unknown origin	1
Secondary to oesophageal varices	1
Unknown	7
Total	36

There were three deaths recorded in 2018; two from infection and one from cardiac arrhythmia. Since 2007, there are seven patients with unknown cause of death, three of which the patients' case notes were already disposed of by the hospital record department. The remaining four patients has no record of case notes and died at home.

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

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

Table 9.4 shows that the number of new cases and new birth are declining in trend.

9 Pahang

9.2.1 Age

Table 9.5 indicates that patients aged 30 years and below forms 77.12% (337 out of 437 patients) of the thalassaemia cases in Pahang. The eldest patient in Pahang is an 87-years-old diagnosed with HbH disease.

Table 9.5: Distribution of Patients in Pahang by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	26	5.95
5 - 9.9	70	16.02
10 -14.9	62	14.19
15 -19.9	61	13.96
20 - 24.9	61	13.96
25 - 29.9	57	13.04
30 - 34.9	35	8.01
35 - 39.9	22	5.03
40 - 44.9	13	2.97
45 - 49.9	12	2.75
50 - 54.9	5	1.14
55 - 59.9	8	1.83
60 - 64.9	1	0.23
Above 65	4	0.92
Total	437	100.00

Figure 9.2: Distribution of Patients in Pahang by Age Group

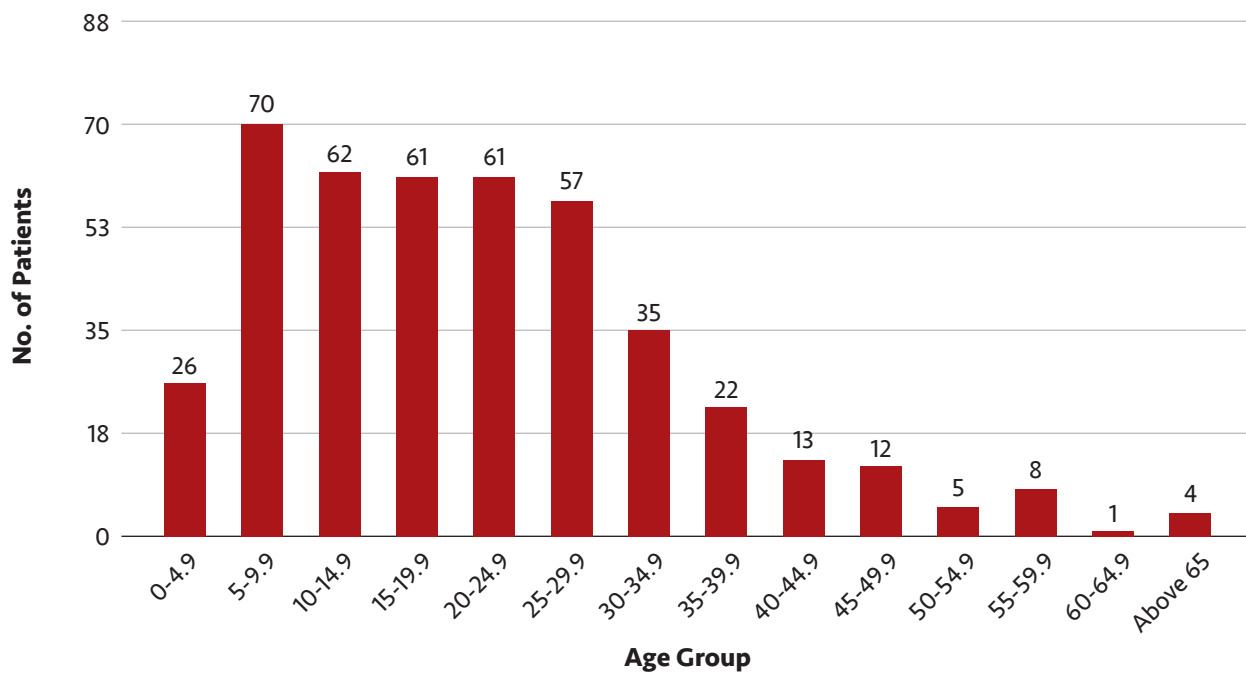


Table 9.6: Distribution of Patients in Pahang According to Diagnosis by Age Group

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	26	Beta Thalassaemia Major	4	0.92
		Beta Thalassaemia Intermedia	3	0.69
		HbE-Beta Thalassaemia	8	1.83
		HbH Disease	7	1.60
		Others	4	0.92
5 - 9.9	70	Beta Thalassaemia Major	7	1.60
		Beta Thalassaemia Intermedia	4	0.92
		HbE-Beta Thalassaemia	24	5.49
		HbH Disease	19	4.35
		Others	16	3.66
10 -14.9	62	Beta Thalassaemia Major	7	1.60
		Beta Thalassaemia Intermedia	2	0.46
		HbE-Beta Thalassaemia	33	7.55
		HbH Disease	20	4.58
		Others	0	0.00
15 -19.9	61	Beta Thalassaemia Major	8	1.83
		Beta Thalassaemia Intermedia	2	0.46
		HbE-Beta Thalassaemia	32	7.32
		HbH Disease	13	2.97
		Others	6	1.37
20 - 24.9	61	Beta Thalassaemia Major	19	4.35
		Beta Thalassaemia Intermedia	6	1.37
		HbE-Beta Thalassaemia	22	5.03
		HbH Disease	9	2.06
		Others	5	1.14
25 - 29.9	57	Beta Thalassaemia Major	17	3.89
		Beta Thalassaemia Intermedia	5	1.14
		HbE-Beta Thalassaemia	27	6.18
		HbH Disease	6	1.37
		Others	2	0.46
30 - 34.9	35	Beta Thalassaemia Major	7	1.60
		Beta Thalassaemia Intermedia	3	0.69
		HbE-Beta Thalassaemia	15	3.43
		HbH Disease	7	1.60
		Others	3	0.69

9 Pahang

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	22	Beta Thalassaemia Major	4	0.92
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	7	1.60
		HbH Disease	5	1.14
		Others	6	1.37
40 - 44.9	13	Beta Thalassaemia Major	3	0.69
		Beta Thalassaemia Intermedia	1	0.23
		HbE-Beta Thalassaemia	7	1.60
		HbH Disease	2	0.46
		Others	0	0.00
45 - 49.9	12	Beta Thalassaemia Major	3	0.69
		Beta Thalassaemia Intermedia	2	0.46
		HbE-Beta Thalassaemia	1	0.23
		HbH Disease	4	0.92
		Others	2	0.46
50 - 54.9	5	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.23
		HbE-Beta Thalassaemia	3	0.69
		HbH Disease	1	0.23
		Others	0	0.00
55 - 59.9	8	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	3	0.69
		HbE-Beta Thalassaemia	2	0.46
		HbH Disease	3	0.69
		Others	0	0.00
60 - 64.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.23
		Others	0	0.00
Above 65	4	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	1	0.23
		HbH Disease	3	0.69
		Others	0	0.00
Total			437	100.00

Thirty-four out of 79 (43.04%) patients with beta thalassaemia major are above 25 years old.

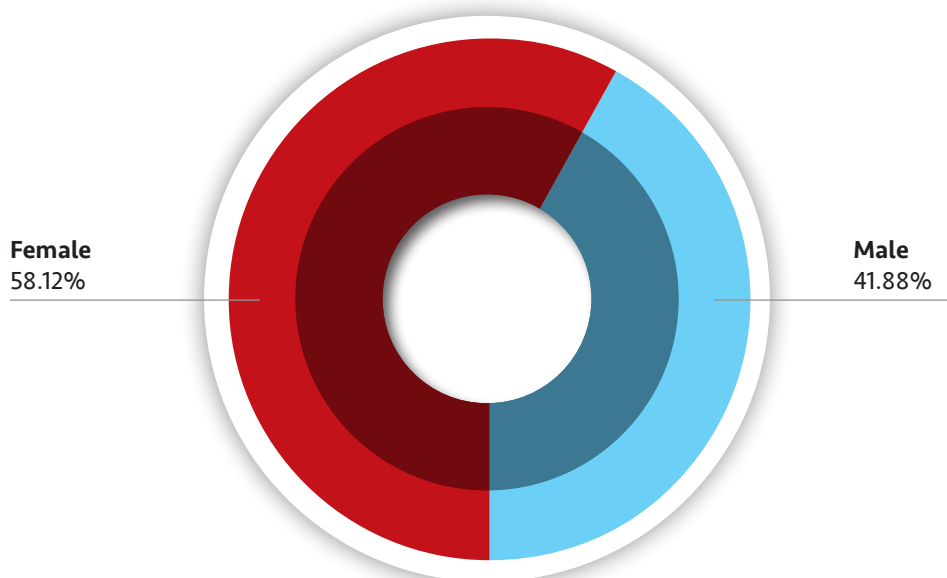
9.2.2. Gender

Table 9.7 shows that female patients dominate the population of thalassaemia patients in Pahang, comprising 58.12% of the total number of patients (254 patients), whereas male patients form 41.88% (183 patients).

Table 9.7: Distribution of Patients in Pahang by Gender

Centre	Male		Female	
	No.	%	No.	%
Tengku Ampuan Afzan, Kuantan (Hospital with specialist)	99	22.65	156	35.70
Sultan Haji Ahmad Shah, Temerloh (Hospital with specialist)	60	13.73	69	15.79
Kuala Lipis (Hospital with specialist)	8	1.83	6	1.37
Pekan (Hospital with specialist)	6	1.37	7	1.60
Raub (Covered by HOSHAS)	1	0.23	5	1.14
Bentong (Covered by HOSHAS)	5	1.14	5	1.14
Jerantut (Covered by HOSHAS)	2	0.46	5	1.14
Jengka (Covered by HOSHAS)	0	0.00	0	0.00
Muadzam Shah, Rompin (Covered by HTAA / Pekan)	2	0.46	1	0.23
Sultanah Hajjah Kalsom, Cameron Highlands (Covered by Ipoh)	0	0.00	0	0.00
Total	183	41.88	254	58.12

Figure 9.3: Distribution of Patients in Pahang by Gender



9 Pahang

9.2.3 Ethnic Group

Majority of thalassaemia patients in Pahang are of Malay descent (393 patients, 89.93%), followed by Chinese (31 patients, 7.09%) and other ethnicities (13 patients, 2.97%).

Table 9.8: Distribution of Patients in Pahang by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	393	89.93
Chinese	31	7.09
Others	13	2.97
Total	437	100

Figure 9.4: Distribution of Patients in Pahang by Ethnic Group

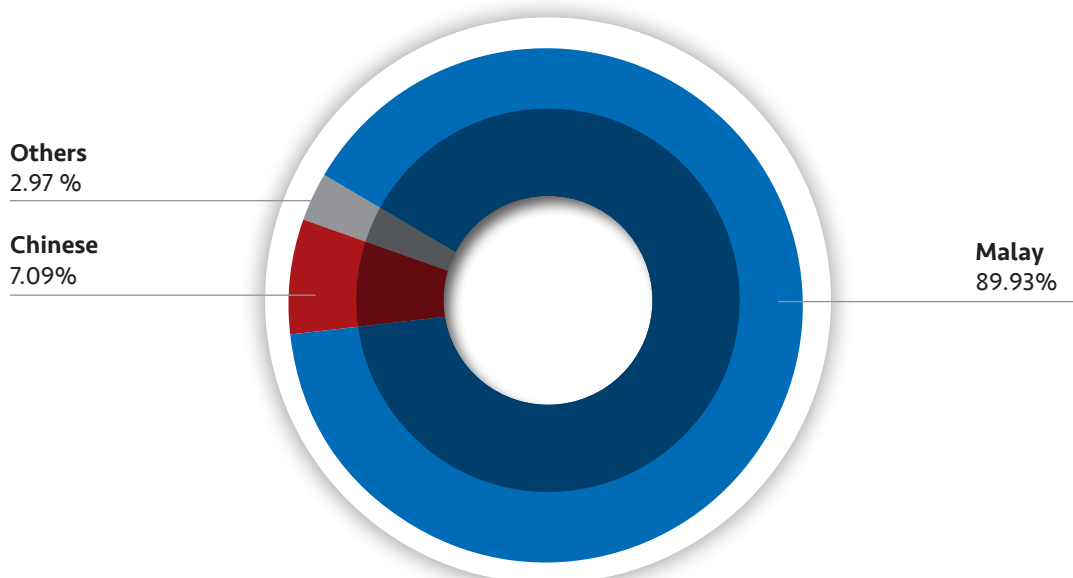


Table 9.9: Distribution of Patients in Pahang According to Ethnic Group by Centre

Centre	Total	Malay		Chinese		Indian		Others	
		No.	%	No.	%	No.	%	No.	%
Tengku Ampuan Afzan, Kuantan	255	234	53.55	18	4.12	0	0.00	3	0.69
Sultan Haji Ahmad Shah, Temerloh	129	115	26.32	7	1.60	0	0.00	7	1.60
Kuala Lipis	14	13	2.97	0	0.00	0	0.00	1	0.23
Pekan	13	12	2.75	0	0.00	0	0.00	1	0.23
Raub	6	5	1.14	1	0.23	0	0.00	0	0.00
Bentong	10	6	1.37	3	0.69	0	0.00	1	0.23
Jerantut	7	6	1.37	1	0.23	0	0.00	0	0.00
Jengka	0	0	0.00	0	0.00	0	0.00	0	0.00
Muadzam Shah, Rompin	3	2	0.46	1	0.23	0	0.00	0	0.00
Total	437	393	89.93	31	7.09	0	0.00	13	2.97

9.3 Diagnosis

The diagnosis with the highest number of patients in Pahang is HbE-beta thalassaemia (182 patients, 41.65%), followed by HbH disease (103 patients, 23.57%), beta thalassaemia major (79 patients, 18.08%), beta thalassaemia intermedia (32 patients, 7.32%) and other diagnoses (41 patients, 9.38%).

Table 9.10: Distribution of Patients in Pahang by Diagnosis

Diagnosis	No. of Patients	Percentage (%)
Beta Thalassaemia Major	79	18.08
Beta Thalassaemia Intermedia	32	7.32
HbE-Beta Thalassaemia	182	41.65
HbH Disease	103	23.57
Others	41	9.38
Total	437	100.00

Figure 9.5: Distribution of Patients in Pahang by Diagnosis

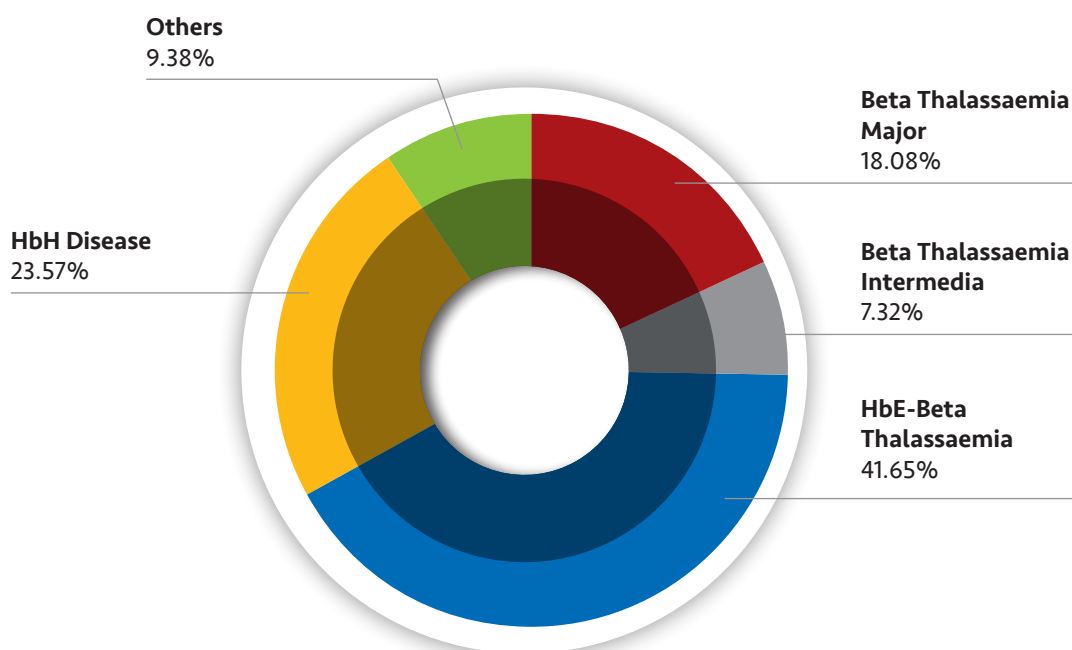


Table 9.11: Distribution of Patients in Pahang According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Tengku Ampuan Afzan, Kuantan	32	7.32	18	4.12	119	27.23	62	14.19	24	5.49
Sultan Haji Ahmad Shah, Temerloh	27	6.18	13	2.97	51	11.67	28	6.41	10	2.29
Kuala Lipis	9	2.06	1	0.23	3	0.69	0	0.00	1	0.23
Pekan	0	0.00	0	0.00	2	0.46	7	1.60	4	0.92
Raub	3	0.69	0	0.00	1	0.23	1	0.23	1	0.23
Bentong	2	0.46	0	0.00	4	0.92	4	0.92	0	0.00
Jerantut	5	1.14	0	0.00	1	0.23	1	0.23	0	0.00
Jengka	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Muadzam Shah, Rompin	1	0.23	0	0.00	1	0.23	0	0.00	1	0.23
Sultanah Hajjah Kalsom, Cameron Highlands	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	79	18.08	32	7.32	182	41.65	103	23.57	41	9.38

Table 9.12: Distribution of Patients in Pahang According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	79	Malay	70	16.02
		Chinese	8	1.83
		Indian	0	0.00
		Others	1	0.23
Beta Thalassaemia Major	32	Malay	31	7.09
		Chinese	1	0.23
		Indian	0	0.00
		Others	0	0.00
HbE-Beta Thalassaemia	182	Malay	169	38.67
		Chinese	8	1.83
		Indian	0	0.00
		Others	5	1.14
HbH Disease	103	Malay	91	20.82
		Chinese	10	2.29
		Indian	0	0.00
		Others	2	0.46
Others	41	Malay	32	7.32
		Chinese	4	0.92
		Indian	0	0.00
		Others	5	1.14
Total			437	100.00

Figure 9.6: Distribution of Alpha Thalassaemia Patients in Pahang by Ethnicity

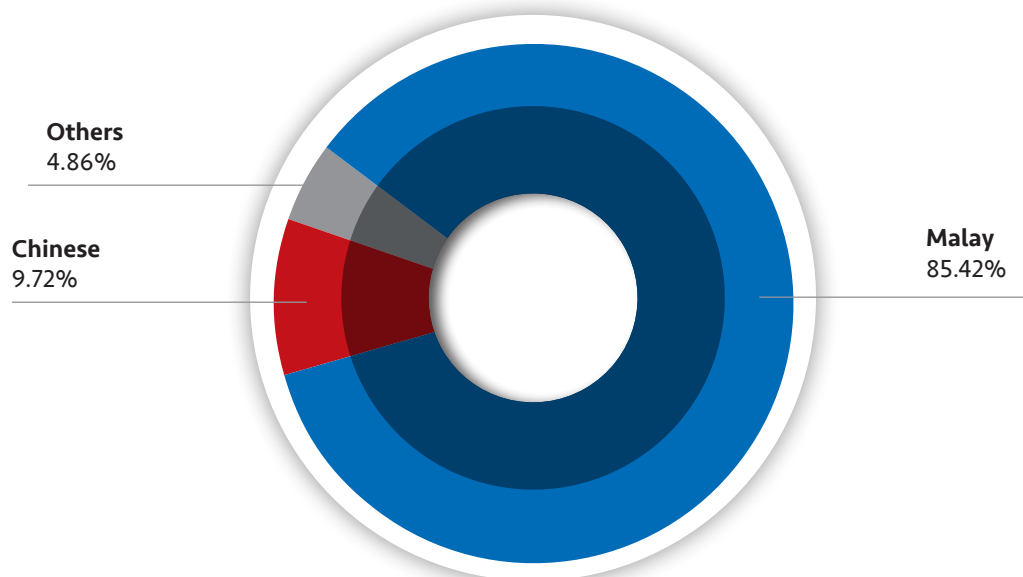
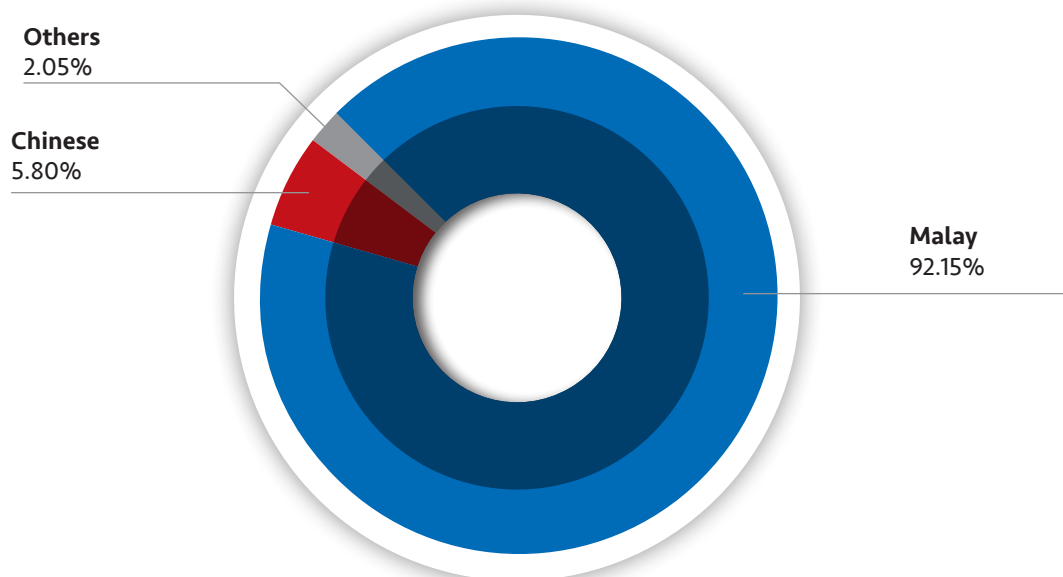


Figure 9.7: Distribution of Beta Thalassaemia Patients in Pahang by Ethnicity



9.4. Treatment

9.4.1 Iron Chelation Therapy

Based on Table 9.13, a total of 267 patients received iron chelating agents. Thirty-three patients (12.36%) received DFO, 83 patients (31.09%) received DFP, 77 patients (28.84%) received DFX, and 55 patients (20.60%) received a combination of DFO and DFP, 5 patients (1.87 %) received a combination of DFP and DFX, and 14 patients (5.24%) received a combination of DFO and DFX. A combination of all three iron chelators (DFO, DFP and DFX) has yet to be recorded in Pahang.

Table 9.13: Distribution of Patients in Pahang by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	33	12.36
DFP only	83	31.09
DFX only	77	28.84
DFO + DFP	55	20.60
DFP + DFX	5	1.87
DFO + DFX	14	5.24
DFO + DFP + DFX	0	0.00
Total	267	100.00

Table 9.14: Distribution of Patients in Pahang According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Tengku Ampuan Afzan, Kuantan (Hospital with specialist)	154	DFO only	17	6.37
		DFP only	53	19.85
		DFX only	39	14.61
		DFO + DFP	33	12.36
		DFP + DFX	2	0.75
		DFO + DFX	10	3.75
		DFO + DFP + DFX	0	0.00
Hospital Sultan Haji Ahmad Shah, Temerloh (Hospital with specialist)	89	DFO only	15	5.62
		DFP only	22	8.24
		DFX only	28	10.49
		DFO + DFP	18	6.74
		DFP + DFX	3	1.12
		DFO + DFX	3	1.12
		DFO + DFP + DFX	0	0.00
Hospital Kuala Lipis (Hospital with specialist)	11	DFO only	1	0.37
		DFP only	5	1.87
		DFX only	5	1.87
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Pekan (Hospital with specialist)	1	DFO only	0	0.00
		DFP only	0	0.00
		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

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Raub (Covered by HOSHAS)	2	DFO only	0	0.00
		DFP only	1	0.37
		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 Bentong (Covered by HOSHAS)	5	DFO only	0	0.00
		DFP only	1	0.37
		DFX only	3	1.12
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.37
		DFO + DFP + DFX	0	0.00
Hospital Jerantut (Covered by HOSHAS)	5	DFO only	0	0.00
		DFP only	1	0.37
		DFX only	0	0.00
		DFO + DFP	4	1.50
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Jengka (Covered by HOSHAS)	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Rompin (Covered by HTAA / Pekan)	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			267	100.00

Figure 9.8: Distribution of Patients by Type of Iron Chelator Received

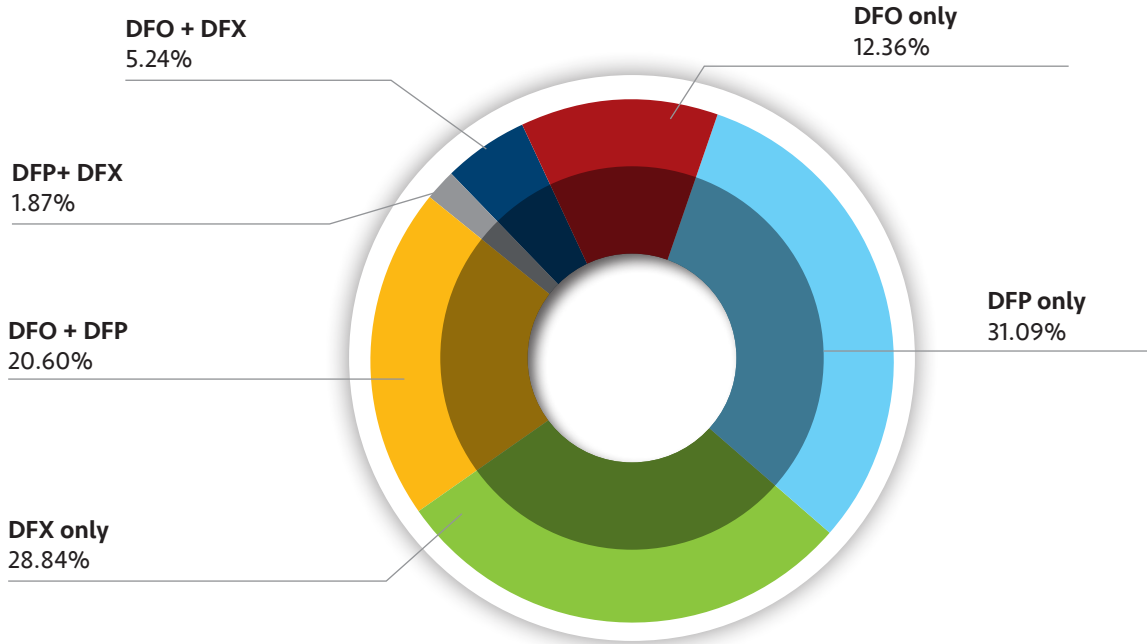


Table 9.15: Distribution of Patients in Pahang According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	5	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	5	1.87
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5-9.9	29	DFO only	1	0.37
		DFP only	0	0.00
		DFX only	26	9.74
		DFO + DFP	2	0.75
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10-14.9	37	DFO only	2	0.75
		DFP only	1	0.37
		DFX only	31	11.61
		DFO + DFP	3	1.12
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
15-19.9	40	DFO only	7	2.62
		DFP only	8	3.00
		DFX only	9	3.37
		DFO + DFP	13	4.87
		DFP + DFX	1	0.37
		DFO + DFX	2	0.75
		DFO + DFP + DFX	0	0.00
20-24.9	48	DFO only	10	3.75
		DFP only	19	7.12
		DFX only	0	0.00
		DFO + DFP	15	5.62
		DFP + DFX	1	0.37
		DFO + DFX	3	1.12
		DFO + DFP + DFX	0	0.00
25-29.9	41	DFO only	5	1.87
		DFP only	18	6.74
		DFX only	4	1.50
		DFO + DFP	8	3.00
		DFP + DFX	0	0.00
		DFO + DFX	6	2.25
		DFO + DFP + DFX	0	0.00
30-34.9	25	DFO only	4	1.50
		DFP only	10	3.75
		DFX only	2	0.75
		DFO + DFP	6	2.25
		DFP + DFX	1	0.37
		DFO + DFX	2	0.75
		DFO + DFP + DFX	0	0.00
35-39.9	11	DFO only	1	0.37
		DFP only	6	2.25
		DFX only	0	0.00
		DFO + DFP	4	1.50
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

9 Pahang

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
40-44.9	12	DFO only	1	0.37
		DFP only	7	2.62
		DFX only	0	0.00
		DFO + DFP	2	0.75
		DFP + DFX	1	0.37
		DFO + DFX	1	0.37
		DFO + DFP + DFX	0	0.00
45-49.9	8	DFO only	1	0.37
		DFP only	6	2.25
		DFX only	0	0.00
		DFO + DFP	1	0.37
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50-54.9	4	DFO only	0	0.00
		DFP only	3	1.12
		DFX only	0	0.00
		DFO + DFP	1	0.37
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	5	DFO only	1	0.37
		DFP only	3	1.12
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	1	0.37
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60-64.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Above 65	2	DFO only	0	0.00
		DFP only	2	0.75
		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

9.4.2 Serum Ferritin Level

There are 152 patients in Pahang who have their serum ferritin level measured in 2018. Table 9.16 shows the distribution of patients by the most recent serum ferritin level in each centre in Pahang. Ninety-eight patients (64.47%) have a serum ferritin level below 2499 ng/mL and 54 patients (35.52%) have a serum ferritin level above 2500 ng/mL. Only 29 patients (19.08%) achieve a serum ferritin level below 1000 ng/mL.

Table 9.16: Distribution of Patients in Pahang According to Most Recent Serum Ferritin Level by Centre

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
HTAA, Kuantan	93	16	10.53	48	31.58	13	8.55	14	9.21	2	1.32
Sultan Haji Ahmad Shah	49	9	5.92	17	11.18	12	7.89	9	5.92	2	1.32
Kuala Lipis	5	2	1.32	1	0.66	2	1.32	0	0.00	0	0.00
Pekan	2	1	0.66	1	0.66	0	0.00	0	0.00	0	0.00
Raub	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Bentong	1	0	0.00	1	0.66	0	0.00	0	0.00	0	0.00
Jerantut	2	1	0.66	1	0.66	0	0.00	0	0.00	0	0.00
Jengka	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Muadzam Shah, Rompin	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Total	152	29	19.08	69	45.39	27	17.76	23	15.13	4	2.63

9.5 Observation and Comments

1. Many older patients (20 years old and above) have high serum ferritin levels and are not well-chelated, as the chelating drugs were not available until much later in 2005 or 2006. There are also compliance issues with DFO.
2. The diagnosis of beta thalassaemia major in some older patients (age range 25-44.9 years old) were probably incorrect and require DNA analysis, as these patients are surviving and receiving regular transfusions without chelating therapy.
3. In the "Others" group of diagnosis, some patients were diagnosed with alpha thalassaemia by Hb analysis. DNA analysis is needed to confirm the diagnosis.
4. 30.72% (90 out of 293) of patients with beta thalassaemia who receive regular transfusion (10 transfusions per year) have had splenectomy performed.
5. Overall, data cleaning is required.
6. Cause of death due to infections may not be accurate as no organisms were specified.



9 Pahang

9.5.1 Recommendations

1. A standardised chart of thalassaemia and patient diary can be introduced across the state and be made available for every thalassaemia patient. The chart is to be used in continuity. By implementing this step, data can be easily collected by the RA.
2. To conduct regular training on thalassaemia for medical staff in Pahang to improve diagnosis, management and outcome due to high turnover of doctors.
3. District hospitals with smaller numbers of thalassaemia patients may train dedicated staff nurses to assist in updating the MTR.
4. The cause of death of thalassaemia patients should be looked into thoroughly, especially involving infections to improve accuracy.

10 Perak

10.1 Introduction

Perak is one of the 13 states in Malaysia, the fourth largest state in the country and the second largest state in Peninsular Malaysia. It has an estimated population of 2.548 million people (according to the Department of Statistics Malaysia 2017). Perak is divided into ten administrative districts, namely Hulu Perak, Larut & Matang, Kerian, Kuala Kangsar, Kinta, Kampar, Perak Tengah, Manjung, Hilir Perak, and Batang Padang. Hospital facilities are provided for each district so patients may receive treatment conveniently. Thalassaemia patients are only treated in Hospital Raja Permaisuri Bainun (Ipoh), Hospital Taiping, Hospital Teluk Intan, Hospital Manjung, Hospital Kuala Kangsar, Hospital Slim River and Hospital Gerik. Furthermore, Hospital Raja Permaisuri Bainun is categorised as the main hospital centre in Perak.

10.2 Patient Demographics

There are 564 thalassaemia patients in Perak. These patients receive transfusions and care in the seven hospitals listed in Table 10.1. Most of these hospitals carry out transfusions in their Ambulatory Care Centre (ACC); for hospitals with no ACC, the transfusions are carried out in the wards. Table 10.1 also shows that most thalassaemia patients receive treatment at Hospital Raja Permaisuri Bainun with 340 patients (60.28%).

Table 10.1: Distribution of Patients in Perak by Centre

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
Hospital Kuala Kangsar	17	3.01
Hospital Raja Permaisuri Bainun	340	60.28
Hospital Seri Manjung	36	6.38
Hospital Slim River	8	1.42
Hospital Taiping	101	17.91
Hospital Teluk Intan	50	8.87
Hospital Gerik	12	2.13
Total	564	100.00

Figure 10.1: Distribution of Patients in Perak by Centre

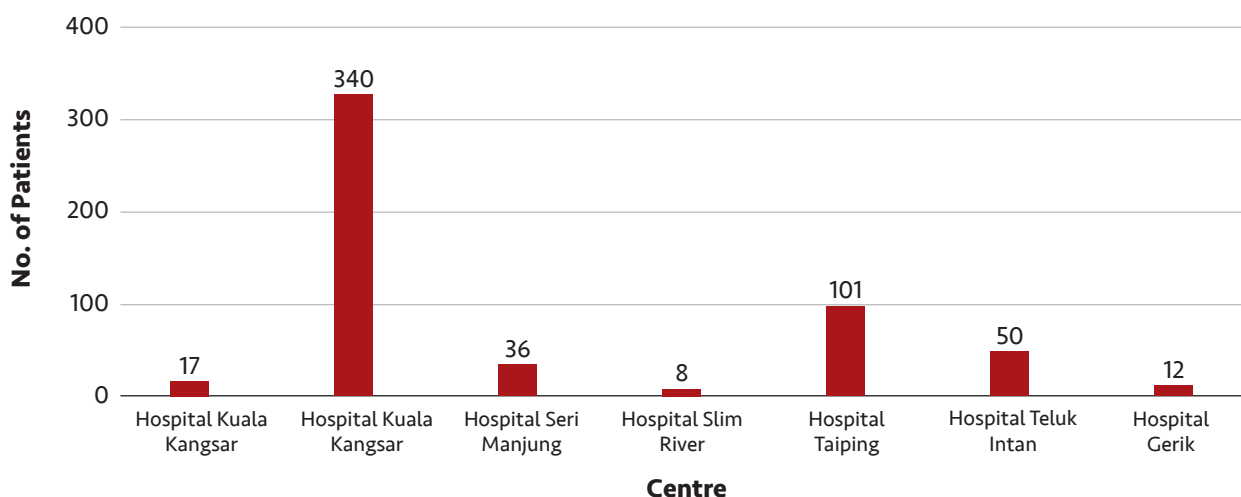


Table 10.2: Distribution of Patients in Perak by Vital Status

Vital Status	No. of Patients
Alive	523
Cumulative Reported Cured by Stem Cell Therapy (2008-2017)	8
Lost to Follow-up	33
Total	564
• Transfusion-dependent thalassaemia (TDT)	246
• Non-transfusion-dependent thalassaemia (NTDT)	318
Deaths in 2018	5
Cumulative Reported Deaths	44

Table 10.3: Cumulative Causes of Death Since 2007 in Perak

Cause of Death	No. of Patients
Cardiac Cause	22
Endocrine Complication	2
Infection	6
Others	13
Unknown*	1
Total	44

*Missing data.

10.2.1 Age

Based on Table 10.4, 43.26% of the thalassaemia patients in Perak are 20 years old and below, and are from the paediatric and adolescent age groups. Patients above 20 years old account for 56.74% of the thalassaemics in Perak.

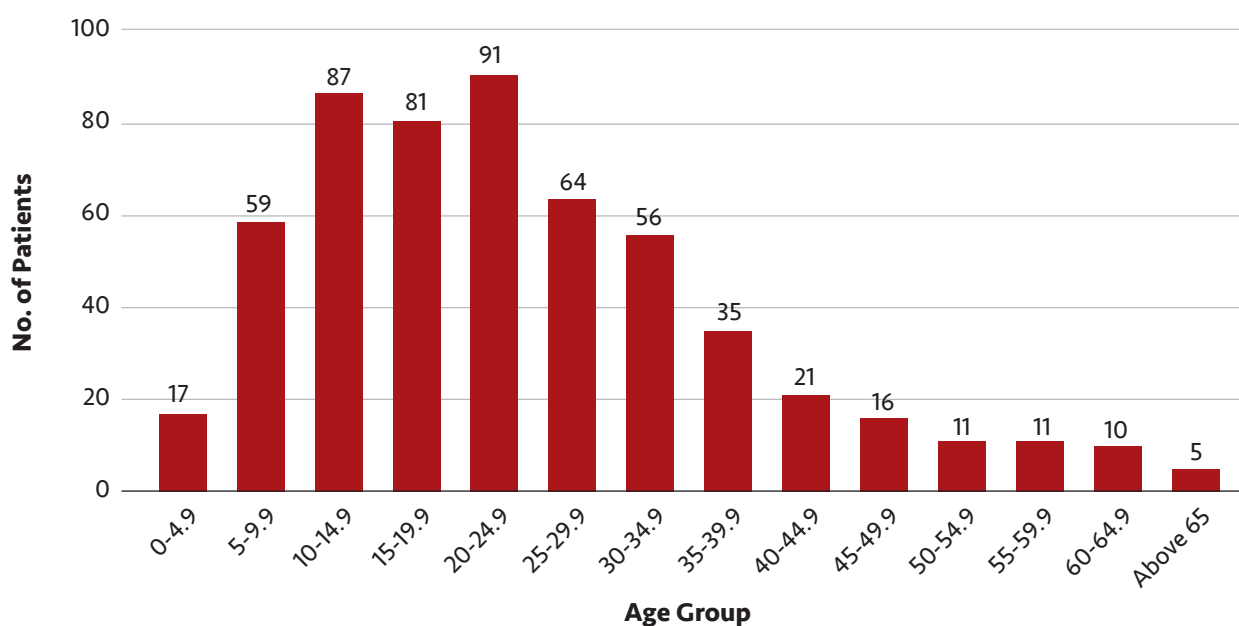
Table 10.4: Distribution of Patients in Perak by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	17	3.01
5 - 9.9	59	10.46
10 -14.9	87	15.43
15 -19.9	81	14.36
20 - 24.9	91	16.13
25 - 29.9	64	11.35
30 - 34.9	56	9.93
35 - 39.9	35	6.21
40 - 44.9	21	3.72
45 - 49.9	16	2.84
50 - 54.9	11	1.95
55 - 59.9	11	1.95
60 - 64.9	10	1.77
Above 65	5	0.89
Total	564	100.00

161

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Figure 10.2: Distribution of Patients in Perak by Age Group



As seen in Figure 10.2, the majority of thalassaemia patients in Perak are at the second and third decade of life.

Table 10.5: Distribution of Patients in Perak According to Diagnosis by Age Group

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	17	Beta Thalassaemia Major	3	0.53
		Beta Thalassaemia Intermedia	2	0.35
		HbE-Beta Thalassaemia	10	1.77
		HbH Disease	1	0.18
		Others	1	0.18
5 - 9.9	59	Beta Thalassaemia Major	11	1.95
		Beta Thalassaemia Intermedia	3	0.53
		HbE-Beta Thalassaemia	26	4.61
		HbH Disease	11	1.95
		Others	8	1.42
10 -14.9	87	Beta Thalassaemia Major	20	3.55
		Beta Thalassaemia Intermedia	11	1.95
		HbE-Beta Thalassaemia	33	5.85
		HbH Disease	17	3.01
		Others	6	1.06
15 -19.9	81	Beta Thalassaemia Major	17	3.01
		Beta Thalassaemia Intermedia	9	1.60
		HbE-Beta Thalassaemia	41	7.27
		HbH Disease	12	2.13
		Others	2	0.35
20 - 24.9	91	Beta Thalassaemia Major	32	5.67
		Beta Thalassaemia Intermedia	9	1.60
		HbE-Beta Thalassaemia	34	6.03
		HbH Disease	15	2.66
		Others	1	0.18
25 - 29.9	64	Beta Thalassaemia Major	10	1.77
		Beta Thalassaemia Intermedia	3	0.53
		HbE-Beta Thalassaemia	30	5.32
		HbH Disease	17	3.01
		Others	4	0.71
30 - 34.9	56	Beta Thalassaemia Major	15	2.66
		Beta Thalassaemia Intermedia	4	0.71
		HbE-Beta Thalassaemia	25	4.43
		HbH Disease	5	0.89
		Others	7	1.24

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	35	Beta Thalassaemia Major	6	1.06
		Beta Thalassaemia Intermedia	3	0.53
		HbE-Beta Thalassaemia	11	1.95
		HbH Disease	10	1.77
		Others	5	0.89
40 - 44.9	21	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	0.35
		HbE-Beta Thalassaemia	6	1.06
		HbH Disease	9	1.60
		Others	4	0.71
45 - 49.9	16	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	8	1.42
		HbH Disease	4	0.71
		Others	4	0.71
50 - 54.9	11	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	5	0.89
		HbH Disease	4	0.71
		Others	2	0.35
55 - 59.9	11	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	0.35
		HbE-Beta Thalassaemia	4	0.71
		HbH Disease	4	0.71
		Others	1	0.18
60 - 64.9	10	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.18
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	5	0.89
		Others	4	0.71
Above 65	5	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	0.35
		HbE-Beta Thalassaemia	1	0.18
		HbH Disease	1	0.18
		Others	1	0.18
Total			564	100.00

10.2.2 Gender

Table 10.6 shows the distribution of thalassaemia patients in Perak by gender. Male and female patients number at 291 (51.60%) and 273 (48.40%), respectively.

Figure 10.3: Distribution of Patients in Perak by Gender

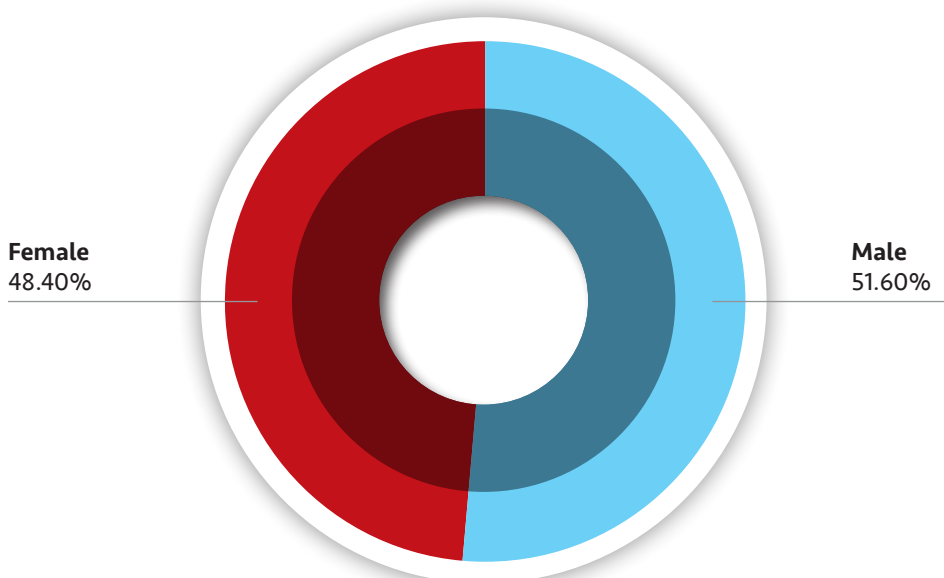


Table 10.6: Distribution of Patients in Perak by Gender

Centre	Male		Female	
	No.	%	No.	%
Hospital Kuala Kangsar	10	1.77	7	1.24
Hospital Raja Permaisuri Bainun	161	28.55	179	31.74
Hospital Seri Manjung	20	3.55	16	2.84
Hospital Slim River	5	0.89	3	0.53
Hospital Taiping	56	9.93	45	7.98
Hospital Teluk Intan	33	5.85	17	3.01
Hospital Gerik	6	1.06	6	1.06
Total	291	51.60	273	48.40

10.2.3 Ethnic Group

Patients from the Malay ethnic group comprises the largest cohort of thalassaemics in Perak with 421 patients (74.65%), followed by the Chinese with 113 patients (20.04%). Other ethnic groups contributed to only a minor proportion of total number of thalassaemics in Perak; this includes Indians with 9 patients (1.60%), Kadazan-Dusun with 1 patient (0.18%), Orang Asli with 18 patients (3.19%) and other ethnic groups with 2 patients (0.35%).

Table 10.7: Distribution of Patients in Perak by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	421	74.65
Chinese	113	20.04
Indian	9	1.60
Orang Asli	18	3.19
Kadazan-Dusun	1	0.18
Others	2	0.35
Total	564	100.00

Figure 10.4: Distribution of Patients in Perak by Ethnic Group

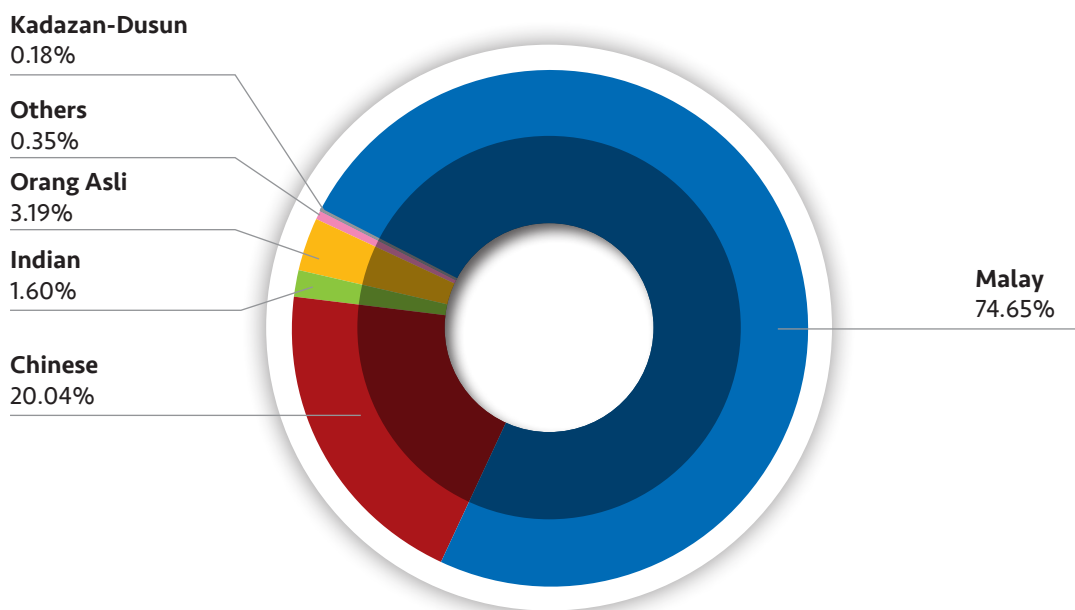


Table 10.8: Distribution of Patients in Perak According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Kadazan-Dusun		Orang Asli		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
H. Kuala Kangsar	16	2.84	1	0.18	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Raja Permaisuri Bainun	225	39.89	96	17.02	7	1.24	11	1.95	0	0.00	1	0.18
Hospital Seri Manjung	32	5.67	4	0.71	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Slim river	7	1.24	0	0.00	0	0.00	0	0.00	1	0.18	0	0.00
Hospital Taiping	94	16.67	6	1.06	1	0.18	0	0.00	0	0.00	0	0.00
Hospital Teluk Intan	38	6.74	6	1.06	1	0.18	4	0.71	0	0.00	1	0.18
Hospital Gerik	9	1.60	0	0.00	0	0.00	3	0.53	0	0.00	0	0.00
Total	421	74.65	113	20.04	9	1.60	18	3.19	1	0.18	2	0.35

10.3 Diagnosis

The diagnosis with the highest number of patients in Perak (234 patients, 41.49%) is HbE-beta thalassaemia, followed by beta thalassaemia major with 114 patients (20.21%), HbH disease with 115 patients (20.39%), beta thalassaemia intermedia with 51 patients (9.04%), while the remaining 50 patients (8.87%) have other forms of haemoglobinopathy such as alpha thalassaemia and Hb Adana.

Table 10.9: Distribution of Patients in Perak by Diagnosis

Diagnosis	No. of Patients	Percentage (%)
Beta Thalassaemia Major	114	20.21
Beta Thalassaemia Intermedia	51	9.04
HbE-Beta Thalassaemia	234	41.49
HbH Disease	115	20.39
Others	50	8.87
Total	564	100.00

Figure 10.5: Distribution of Patients in Perak by Diagnosis

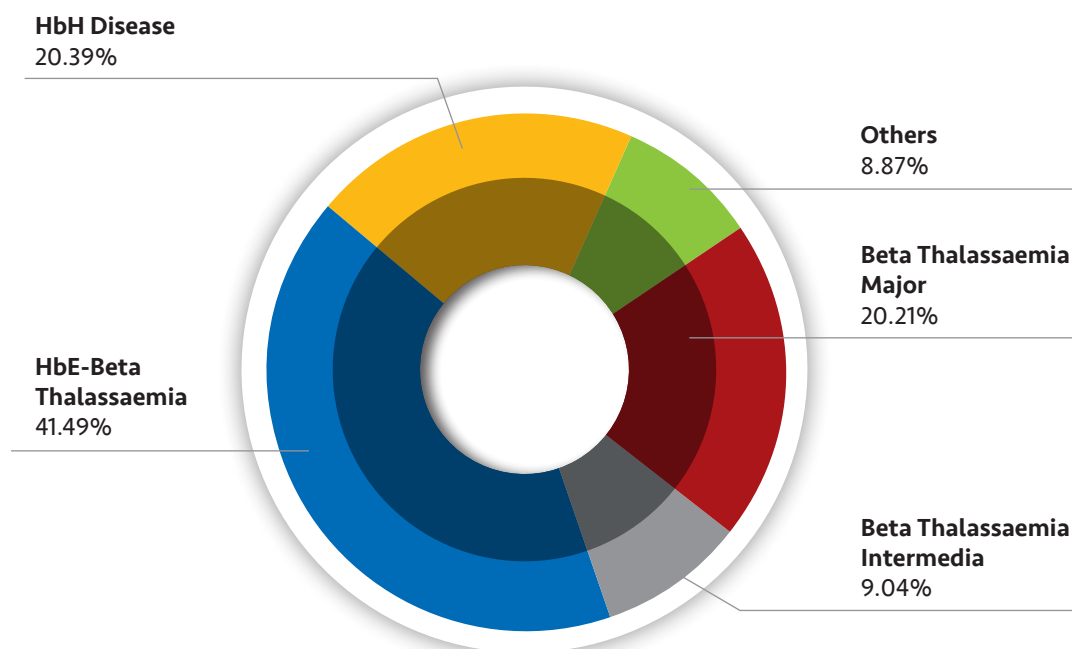


Table 10.10: Distribution of Patients in Perak According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Kuala Kangsar	3	0.53	0	0.00	11	1.95	0	0.00	3	0.53
Hospital Raja Permaisuri Bainun	57	10.11	32	5.67	124	21.99	91	16.13	36	6.38
Hospital Seri Manjung	17	3.01	1	0.18	12	2.13	5	0.89	1	0.18
Hospital Slim River	3	0.53	0	0.00	5	0.89	0	0.00	0	0.00
Hospital Taiping	23	4.08	11	1.95	49	8.69	13	2.30	5	0.89
Hospital Teluk Intan	9	1.60	7	1.24	30	5.32	1	0.18	3	0.53
Hospital Gerik	2	0.35	0	0.00	3	0.53	5	0.89	2	0.35
Total	114	20.21	51	9.04	234	41.49	115	20.39	50	8.87

Table 10.11: Distribution of Patients in Perak According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	114	Malay	74	13.12
		Chinese	37	6.56
		Indian	1	0.18
		Kadazan-Dusun	0	0.00
		Orang Asli	1	0.18
		Others	1	0.18
Beta Thalassaemia Major	51	Malay	39	6.91
		Chinese	10	1.77
		Indian	1	0.18
		Kadazan-Dusun	0	0.00
		Orang Asli	1	0.18
		Others	0	0.00
HbE-Beta Thalassaemia	234	Malay	204	36.17
		Chinese	19	3.37
		Indian	1	0.18
		Kadazan-Dusun	0	0.00
		Orang Asli	9	1.60
		Others	1	0.18
HbH Disease	115	Malay	73	12.94
		Chinese	36	6.38
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Orang Asli	6	1.06
		Others	0	0.00
Others	50	Malay	31	5.50
		Chinese	11	1.95
		Indian	7	1.24
		Kadazan-Dusun	0	0.00
		Orang Asli	1	0.18
		Others	0	0.00
Total			564	100.00

10.4 Treatment

10.4.1 Iron Chelation Therapy

Based on Table 10.12, 329 patients in Perak received iron chelation therapy. Eighty-four patients (25.53%) are receiving DFO therapy only, and 78 patients (23.71%) are receiving oral DFP therapy only. Another 105 patients (31.91%) are on DFX, which is normally reserved for patients who could not tolerate other chelating agents. Furthermore, 49 patients (14.89%) are receiving a combination of DFO and DFP. In addition, patients taking a combination of DFP/DFX and DFO/DFX number at 6 (1.82%) and 4 (1.22%) patients, respectively. Only 3 patients (0.91%) are receiving a DFO/DFP/DFX combination. Nevertheless, a high proportion of patients in Perak (235 patients, 41.67%) have not received iron chelation therapy.

Table 10.12: Distribution of Patients in Perak by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	84	25.53
DFP only	78	23.71
DFX only	105	31.91
DFO + DFP	49	14.89
DFP + DFX	6	1.82
DFO + DFX	4	1.22
DFO + DFP + DFX	3	0.91
Total	329	100.00

Table 10.13: Distribution of Patients in Perak According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Kuala Kangsar	12	DFO only	4	1.22
		DFP only	3	0.91
		DFX only	3	0.91
		DFO + DFP	2	0.61
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
HRPB, Ipoh	175	DFO only	48	14.59
		DFP only	46	13.98
		DFX only	48	14.59
		DFO + DFP	26	7.90
		DFP + DFX	3	0.91
		DFO + DFX	1	0.30
		DFO + DFP + DFX	3	0.91

10 Perak

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Seri Manjung	28	DFO only	7	2.13
		DFP only	2	0.61
		DFX only	14	4.26
		DFO + DFP	3	0.91
		DFP + DFX	0	0.00
		DFO + DFX	2	0.61
		DFO + DFP + DFX	0	0.00
Hospital Slim River	3	DFO only	3	0.91
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Taiping	73	DFO only	9	2.74
		DFP only	21	6.38
		DFX only	24	7.29
		DFO + DFP	16	4.86
		DFP + DFX	3	0.91
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Teluk Intan	35	DFO only	13	3.95
		DFP only	6	1.82
		DFX only	13	3.95
		DFO + DFP	2	0.61
		DFP + DFX	0	0.00
		DFO + DFX	1	0.30
		DFO + DFP + DFX	0	0.00
Hospital Gerik	3	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	3	0.91
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			329	100.00

Figure 10.6: Distribution of Patients in Perak by Type of Iron Chelator Received

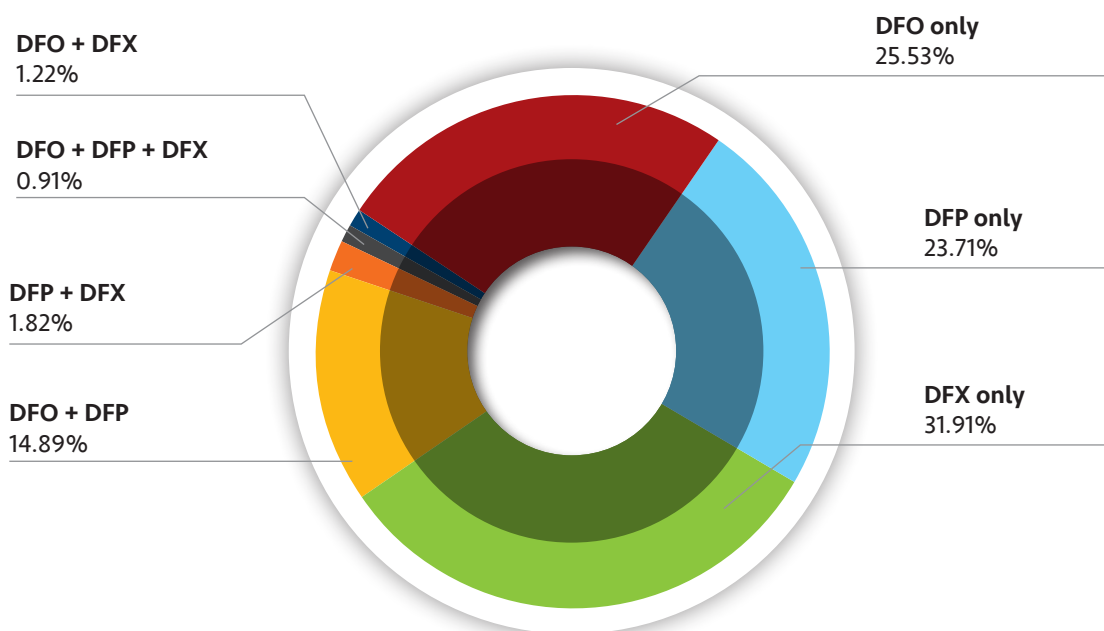


Table 10.14: Distribution of Patients in Perak According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	10	DFO only	0	0.00
		DFP only	2	0.61
		DFX only	7	2.13
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.30
		DFO + DFP + DFX	0	0.00
5-9.9	31	DFO only	6	1.82
		DFP only	2	0.61
		DFX only	23	6.99
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10-14.9	49	DFO only	9	2.74
		DFP only	2	0.61
		DFX only	38	11.55
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

10 Perak

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
15-19.9	51	DFO only	14	4.26
		DFP only	8	2.43
		DFX only	20	6.08
		DFO + DFP	8	2.43
		DFP + DFX	1	0.30
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20-24.9	62	DFO only	20	6.08
		DFP only	11	3.34
		DFX only	9	2.74
		DFO + DFP	17	5.17
		DFP + DFX	3	0.91
		DFO + DFX	1	0.30
		DFO + DFP + DFX	1	0.30
25-29.9	36	DFO only	12	3.65
		DFP only	8	2.43
		DFX only	1	0.30
		DFO + DFP	11	3.34
		DFP + DFX	1	0.30
		DFO + DFX	1	0.30
		DFO + DFP + DFX	2	0.61
30-34.9	42	DFO only	11	3.34
		DFP only	19	5.78
		DFX only	3	0.91
		DFO + DFP	8	2.43
		DFP + DFX	1	0.30
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
35-39.9	15	DFO only	5	1.52
		DFP only	6	1.82
		DFX only	1	0.30
		DFO + DFP	3	0.91
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
40-44.9	8	DFO only	2	0.61
		DFP only	5	1.52
		DFX only	1	0.30
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-49.9	5	DFO only	2	0.61
		DFP only	2	0.61
		DFX only	1	0.30
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50-54.9	4	DFO only	1	0.30
		DFP only	3	0.91
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	7	DFO only	1	0.30
		DFP only	3	0.91
		DFX only	0	0.00
		DFO + DFP	2	0.61
		DFP + DFX	0	0.00
		DFO + DFX	1	0.30
		DFO + DFP + DFX	0	0.00
60-64.9	6	DFO only	0	0.00
		DFP only	5	1.52
		DFX only	1	0.30
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Above 65	3	DFO only	1	0.30
		DFP only	2	0.61
		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			329	100.00

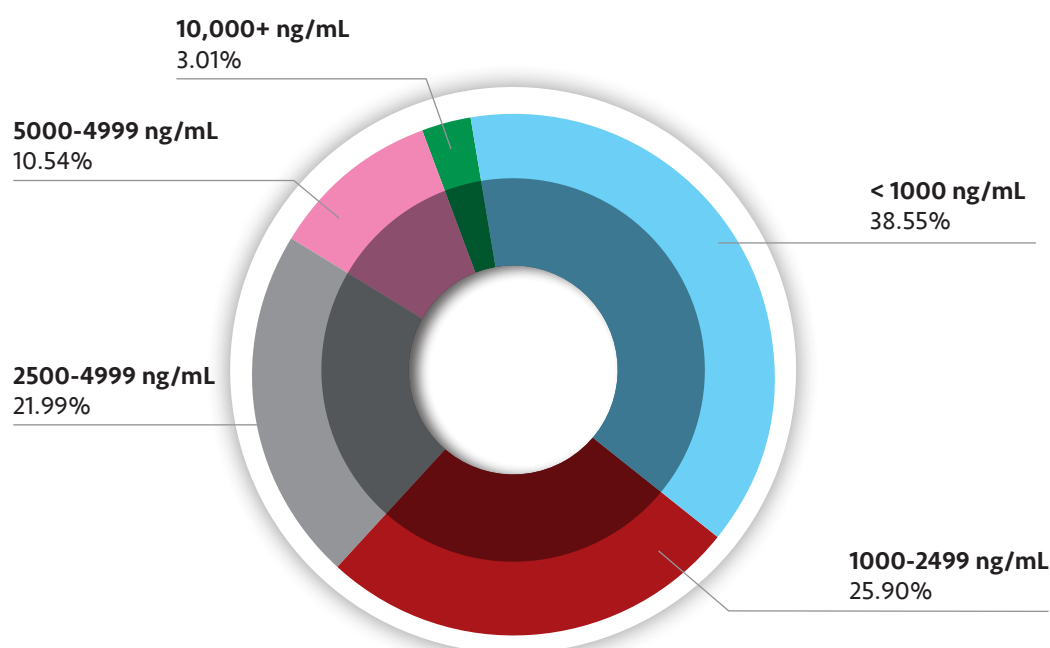
10.4.2 Serum Ferritin Level

Overall, only 332 patients (58.87%) had their most recent ferritin level measured in 2018. About 128 patients (38.55%) have a serum ferritin level lower than 1000 ng/mL, which is ideal. Eighty-six patients (25.90%) have a serum ferritin level between 1000 to 2499 ng/mL. This range is acceptable in preventing end organ damage due to iron overload. This leaves 118 patients (35.54%) with a known serum ferritin level above 2500 ng/mL.

Table 10.15: Distribution of Patients in Perak According to Most Recent Serum Ferritin Level by Centre

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Kuala Kangsar	6	1	0.30	2	0.60	0	0.00	2	0.60	1	0.30
Hospital Raja Permaisuri Bainun	215	97	29.22	57	17.17	44	13.25	16	4.82	1	0.30
Hospital Seri Manjung	16	4	1.20	5	1.51	4	1.20	2	0.60	1	0.30
Hospital Slim River	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Taiping	68	19	5.72	15	4.52	16	4.82	11	3.31	7	2.11
Hospital Teluk Intan	24	7	2.11	7	2.11	7	2.11	3	0.90	0	0.00
Hospital Gerik	3	0	0.00	0	0.00	2	0.60	1	0.30	0	0.00
Total	332	128	38.55	86	25.90	73	21.99	35	10.54	10	3.01

Figure 10.7: Distribution of Patients in Perak by Most Recent Serum Ferritin Level



Only 181 TDT patients (73.58%) in Perak had their most recent ferritin level measured in 2018. About 34 patients (18.78%) have a serum ferritin level ideally below 1000 ng/mL. Fifty-nine patients (32.60%) have a serum ferritin level between 1000 to 2499 ng/mL whereas 88 patients (48.62%) have a known serum ferritin level above 2500 ng/mL.

Table 10.16: Distribution of TDT Patients in Perak According to Most Recent Serum Ferritin Level by Centre

Serum Ferritin Level (ng/mL)	Centre	Total	< 1000		1000-2499		2500-4999		5000-9999		10,000+	
			No.	%	No.	%	No.	%	No.	%	No.	%
	Hospital Kuala Kangsar	3	1	0.55	1	0.55	0	0.00	1	0.55	0	0.00
	Hospital Raja Permaisuri Bainun	112	23	12.71	38	20.99	37	20.44	13	7.18	1	0.55
	Hospital Seri Manjung	8	3	1.66	2	1.10	0	0.00	2	1.10	1	0.55
	Hospital Slim River	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
	Hospital Taiping	38	5	2.76	11	6.08	8	4.42	8	4.42	6	3.31
	Hospital Teluk Intan	17	2	1.10	7	3.87	5	2.76	3	1.66	0	0.00
	Hospital Gerik	3	0	0.00	0	0.00	2	1.10	1	0.55	0	0.00
Total		181	34	18.78	59	32.60	52	28.73	28	15.47	8	4.42

10.5 Observation and Comments

This report gives a brief overview on the number of thalassaemia patients, the type of thalassaemia and the treatment provided to all thalassaemia patients in the state of Perak. Serum ferritin levels below 1000 ng/mL indicate good compliance to iron chelators; however, only 18.78% of patients fall in this category. About 64.46% of patients have an acceptable serum ferritin level lower than 2500 ng/mL. Thus, compliance still remains a major problem in Perak (as shown in Table 10.15). Based on Table 10.16, approximately half of the total number of patients (51.38%) are within the desirable ferritin level. Therefore, we have undertaken several measures to improve the statistic, 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.

Over the last five years, there have been 23 documented new births with thalassaemia in Perak. A total of five deaths were reported in 2018. The deaths were mainly due to cardiac complications, among which was a case of severe pulmonary hypertension, decompensated congestive cardiac failure with cardiac arrhythmia. There was only one mortality due to severe infection.

10.5.1 Recommendations

1. A standardised documentation and charting of the results for all thalassaemia patients and a personal patient diary can be made available for easy access by the treating clinicians all over the state. This practise can also improve the effectiveness of data entry by the RA.
2. To conduct regular training for all healthcare personnel on management of thalassaemia patients.
3. To require staff nurses to record data on patients' date and cause of death.

10.5.2 Conclusions

1. The number of thalassaemia patients in the state of Perak for the year 2018 was reported at 564 patients. Of this number, TDT comprises 246 patients (43.62%) and NTDT comprises patients 318 patients (56.38%).
2. The majority of thalassaemia patients in Perak are adults with 320 patients (56.74%), whereas paediatric patients number at 244 patients (43.26%).
3. The subtypes of thalassaemia found in the state were mainly HbE-beta thalassaemia (41.49%) followed by beta thalassaemia major (20.21%).
4. The ethnic distribution of thalassaemia in Perak reported in the national data registry consists of Malays (74.65%), Chinese (20.04), and other ethnicities (5.32%).
5. The mortalities reported in Perak in 2018 involved four adults and one paediatric patient. The causes of death were cardiac complications and infection. These were also the major causes of death in the national registry data.
6. Total number of patients on chelation therapy in the state of Perak is 329 patients (58.33%). A total of 230 TDT patients (93.5%) are currently receiving chelation therapy. The remaining 6.5% of TDT patients were not on chelation therapy, mainly due to non-compliance to treatment or was lost to follow-up. A total of 99 NTDT patients (31.13%) are also on chelation therapy.
7. The main chelation therapy used was DFX (31.91%), followed by DFO (25.53%). 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 was DFO/DFP.
8. A total of 181 (73.58%) out of 246 TDT patients had their serum ferritin level measured in 2018. Of the 181 patients, 51.38% have a serum ferritin level below 2499 ng/mL.

11 Perlis

11.1 Introduction

Perlis is the smallest state in Malaysia. Perlis has an estimation population of 253,800 people as of in 2018 (Department of Statistics Malaysia). There is only one hospital in Perlis , which is Hospital Tuanku Fauziah (HTF) located in Kangar.

11.2 Patient Demographics

Data analysed were taken from patients with status of either alive, lost to follow-up or cured by transplant, and excludes deceased patients. There are 125 living patients in Perlis. One patient was cured by transplant and 12 patients were deceased.

Table 11.1: Distribution of Patients in Perlis by Centre

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
Hospital Tuanku Fauziah, Kangar	128	100.00
Total	128	100.00

Table 11.2: Distribution of Patients in Perlis by Vital Status

Vital Status	No. Of Patients
Alive	125
Cumulative Reported Cured by Stem Cell	1
Lost to Follow-up	2
Total	128
Deaths in 2018	0
Cumulative Reported Deaths	12

Table 11.3: Cumulative Causes of Death Since 2007 in Perlis

Cause of Death	No. of Patients
Infections	1
Cardiac causes	7
Motor Vehicle Accident	3
Liver disease	1
Total	12

11.2.1 Age

The youngest patient in Perlis is 1 year old and the eldest is 63 years old. Sixty-six patients (51.56%) are below 20 years old. The median age group is 15-19.9 years old.

Table 11.4: Distribution of Patients in Perlis by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	5	3.91
5 - 9.9	12	9.38
10 -14.9	24	18.75
15 -19.9	25	19.53
20 - 24.9	35	27.34
25 - 29.9	16	12.50
30 - 34.9	6	4.69
35 - 39.9	1	0.78
40 - 44.9	0	0.00
45 - 49.9	1	0.78
50 - 54.9	0	0.00
55 - 59.9	1	0.78
60 - 64.9	2	1.56
Above 65	0	0.00
Total	128	100.00

Figure 11.1: Distribution of Patients in Perlis by Age Group

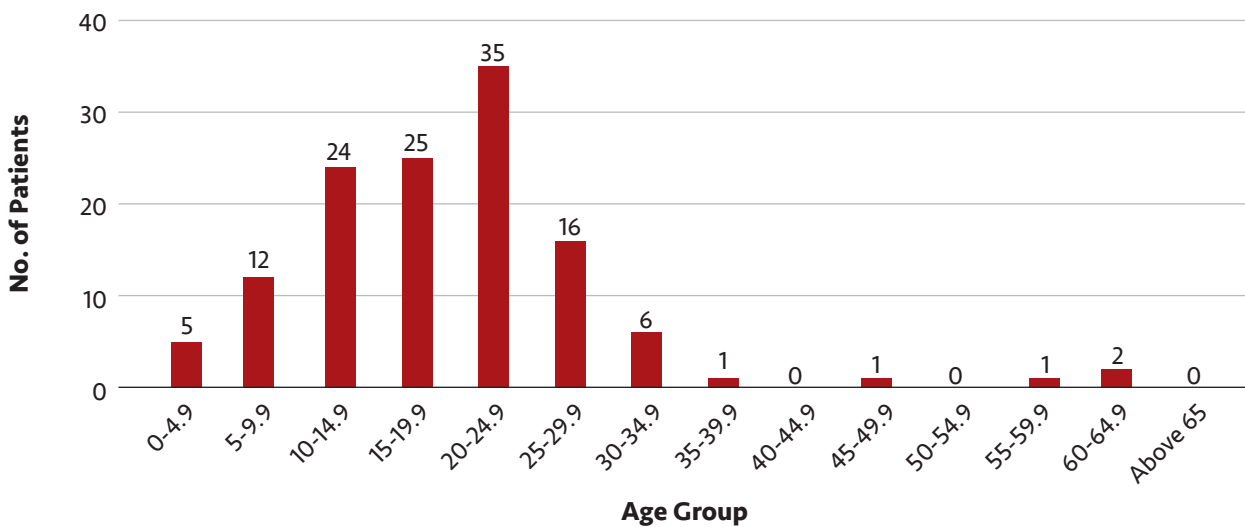


Table 11.5: Distribution of Patients in Perlis According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	5	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	1.56
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	2	1.56
		Others	1	0.78
5 - 9.9	12	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	5	3.91
		HbE-Beta Thalassaemia	4	3.13
		HbH Disease	3	2.34
		Others	0	0.00
10 -14.9	24	Beta Thalassaemia Major	8	6.25
		Beta Thalassaemia Intermedia	3	2.34
		HbE-Beta Thalassaemia	6	4.69
		HbH Disease	4	3.13
		Others	3	2.34
15 -19.9	25	Beta Thalassaemia Major	2	1.56
		Beta Thalassaemia Intermedia	5	3.91
		HbE-Beta Thalassaemia	10	7.81
		HbH Disease	5	3.91
		Others	3	2.34
20 - 24.9	35	Beta Thalassaemia Major	9	7.03
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	17	13.28
		HbH Disease	9	7.03
		Others	0	0.00
25 - 29.9	16	Beta Thalassaemia Major	3	2.34
		Beta Thalassaemia Intermedia	1	0.78
		HbE-Beta Thalassaemia	11	8.59
		HbH Disease	1	0.78
		Others	0	0.00
30 - 34.9	6	Beta Thalassaemia Major	3	2.34
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	3	2.34
		HbH Disease	0	0.00
		Others	0	0.00

11 Perlis

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	1	Beta Thalassaemia Major	1	0.78
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
40 - 44.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
45 - 49.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.0
		HbE-Beta Thalassaemia	1	0.78
		HbH Disease	0	0.00
		Others	0	0.00
50 - 54.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
55 - 59.9	1	Beta Thalassaemia Major	0	0.0
		Beta Thalassaemia Intermedia	1	0.78
		HbE-Beta Thalassaemia	0	0.0
		HbH Disease	0	0.0
		Others	0	0.0
60 - 64.9	2	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	1	0.78
		HbH Disease	0	0.00
		Others	1	0.78
Above 65	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			128	100.00

11.2.2 Gender

Table 11.6 shows the distribution of thalassaemia patients in Perlis by gender. Number of male patients are higher (72 patients, 56.25 %) compared to the female patients (56, 43.75%).

Figure 11.2: Distribution of Patients in Perlis by Gender

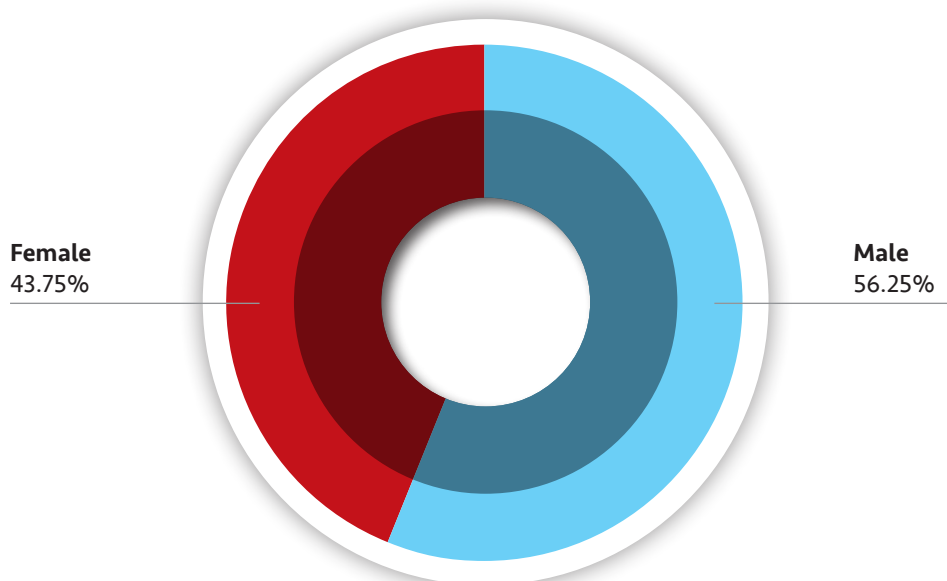


Table 11.6: Distribution of Patients in Perlis by Gender

Centre	Male		Female	
	No.	%	No.	%
HTF, Kangar	72	56.25	56	43.75
Total	72	56.25	56	43.75

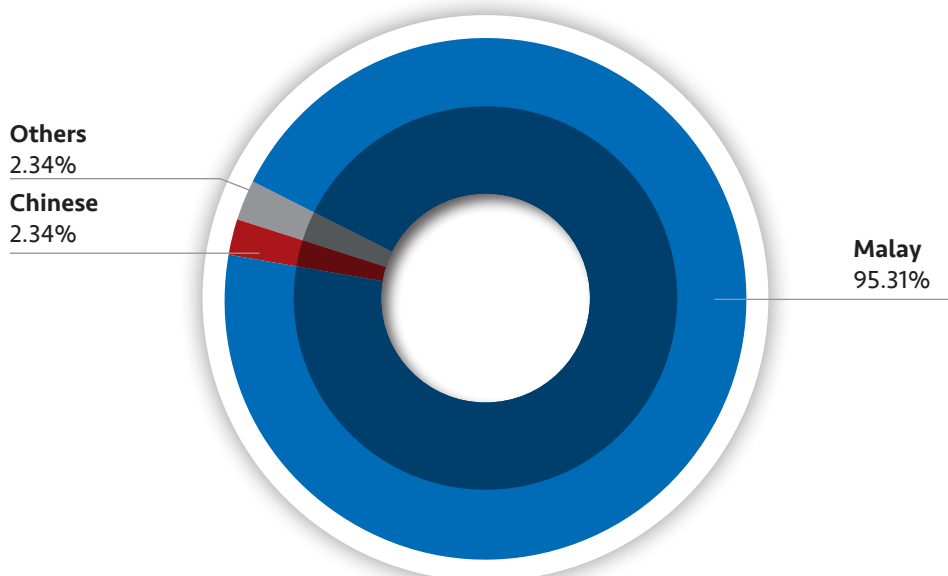
11.2.3 Ethnic Group

As shown in Table 11.7, majority of the thalassaemia patients in Perlis are of Malay descent (122 patients, 95.31%). Three patients (2.34%) are of Chinese descent. The other ethnic groups recorded were Thai with two patients (1.56%) and Murut with one patient (0.78%).

Table 11.7: Distribution of Patients in Perlis by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	122	95.31
Chinese	3	2.34
Others:		
Thai	2	1.56
Murut	1	0.78
Total	128	100

Figure 11.3: Distribution of Patients in Perlis by Ethnic Group



11.3 Diagnosis

The most common diagnosis among patients in Perlis is HbE-beta thalassaemia (53 patients, 41.41%), followed by beta thalassaemia major (26 patients, 20.31%), HbH disease (24 patients, 18.75%) and beta thalassaemia intermedia (17 patients, 13.28%). The remaining 8 patients (6.25%) has other diagnosis (Table 11.8).

Table 11.8: Distribution of Patients in Perlis by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	26	20.31
Beta Thalassaemia Intermedia	17	13.28
HbE-Beta Thalassaemia	53	41.41
HbH Disease	24	18.75
Others	8	6.25
Total	128	100.00

Figure 11.4: Distribution of Patients in Perlis by Diagnosis

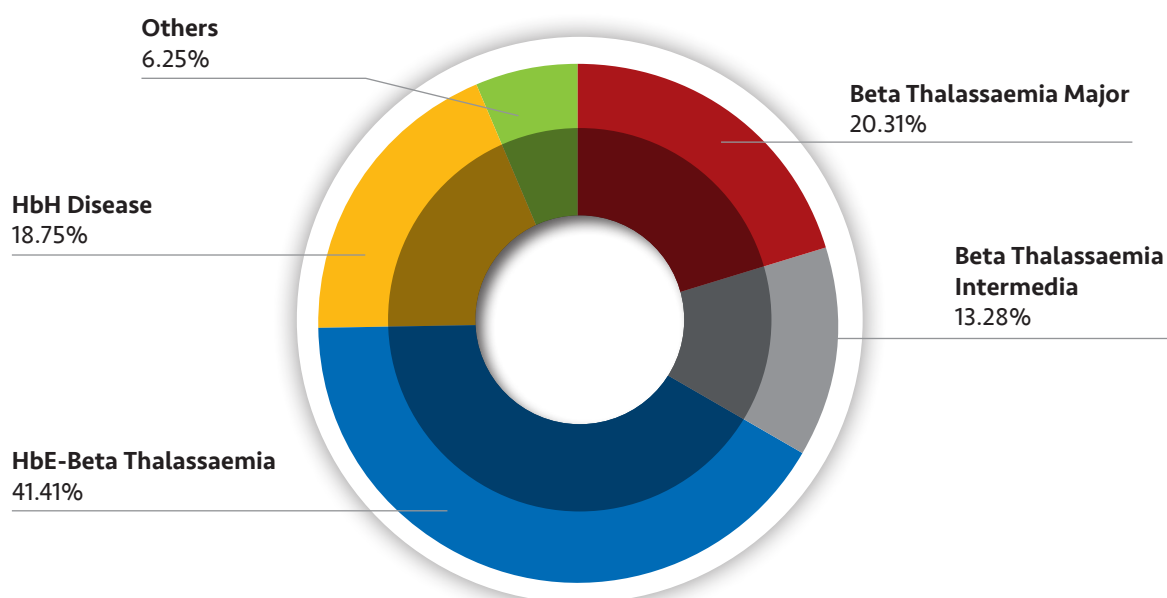


Table 11.9: Distribution of Patients in Perlis According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	26	Malay	25	19.53
		Chinese	1	0.78
		Indian	0	0.00
		Others	0	0.00
Beta Thalassaemia Intermedia	17	Malay	17	13.28
		Chinese	0	0.00
		Indian	0	0.00
		Others	0	0.00
HbE-Beta Thalassaemia	53	Malay	51	39.84
		Chinese	1	0.78
		Indian	0	0.00
		Others	1	0.78
HbH Disease	24	Malay	22	17.19
		Chinese	1	0.78
		Indian	0	0.00
		Others	1	0.78
Others	8	Malay	7	5.47
		Chinese	0	0.00
		Indian	0	0.00
		Others	1	0.78
Total			128	100.00

11.4 Treatment

11.4.1 Iron Chelation Therapy

About 74 patients (57.81%) in Perlis received iron chelation therapy. Sixty-five (87.84%) of the 74 chelated patients are on oral DFX, five patients (6.76%) are on subcutaneous DFO, and two patients (2.70%) each are on oral DFP and on combination therapy.

Table 11.10: Distribution of Patients in Perlis by Type of Iron Chelator Received

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
HTF, Kangar	74	DFO only	5	6.76
		DFP only	2	2.70
		DFX only	65	87.84
		DFO + DFP	2	2.70
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			74	100.00

Figure 11.5: Distribution of Patients in Perlis by Type of Iron Chelator Received

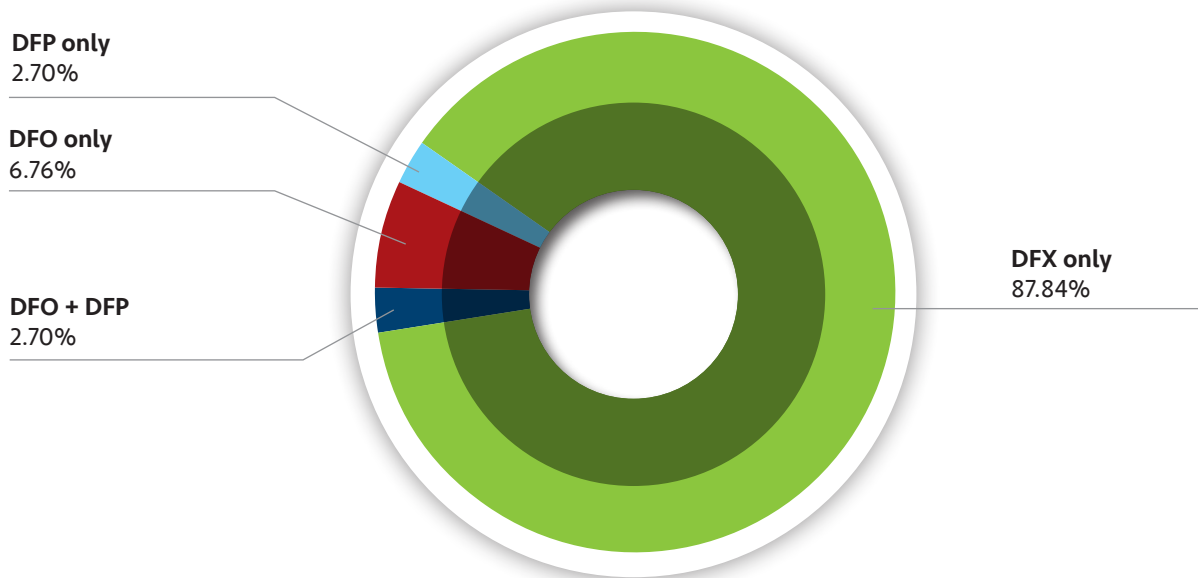


Table 11.11: Distribution of Patients in Perlis According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	0	DFO	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5-9.9	3	DFO	0	0.00
		DFP only	0	0.00
		DFX only	3	4.05
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10-14.9	15	DFO	1	1.35
		DFP only	0	0.00
		DFX only	14	18.92
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15-19.9	15	DFO	1	1.35
		DFP only	0	0.00
		DFX only	14	18.92
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20-24.9	24	DFO	2	2.70
		DFP only	0	0.00
		DFX only	20	27.03
		DFO + DFP	2	2.70
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

11 Perlis

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
25-29.9	9	DFO	1	1.35
		DFP only	0	0.00
		DFX only	8	10.81
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30-34.9	5	DFO	0	0.00
		DFP only	1	1.35
		DFX only	4	5.41
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
35-39.9	1	DFO	0	0.00
		DFP only	0	0.00
		DFX only	1	1.35
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40-44.9	0	DFO	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-49.9	1	DFO	0	0.00
		DFP only	0	0.00
		DFX only	1	1.35
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
50-54.9	0	DFO	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	0	DFO	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60-64.9	1	DFO	0	0.00
		DFP only	1	1.35
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	0	DFO	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			74	100.00

11.4.2 Serum Ferritin Level

There are 33 patients in Perlis who had their serum ferritin level measured. There are 18 patients (54.54%) with serum ferritin level lower than 2500 ng/mL, and 15 patients (45.45%) with moderate to severe iron overload, with serum ferritin level above 2500 ng/mL.

Table 11.12: Distribution of Patients in Perlis According to Most Recent Serum Ferritin Level by Centre

Serum Ferritin Level (ng/mL)	Centre	< 1000		1000-2499		2500-4999		5000-9999		10,000+		
		Total	No.	%	No.	%	No.	%	No.	%	No.	%
	HTF, Kangar	33	5	15.15	13	39.39	10	30.30	5	15.15	0	0.00
Total		33	5	15.15	13	39.39	10	30.30	5	15.15	0	0.00



11 Perlis

11.5 Observation and Comments

Approximately 52% of thalassaemia patients in Perlis are below 20 years old. HbE-beta thalassaemia is the main type diagnosed in Perlis (41.41% of all cases). Sixty-five chelated patients (87.84%) are on oral DFX.

T2* MRI services are available in HTF and is a great help in the assessment of iron overload. However, there is a need for details in the report for validation of data from time to time, to ensure quality care is being provided at all times.

11.5.1 Recommendations

Patients compliance need to be checked regularly and combination therapy is needed in severely iron overloaded patients for better outcomes. There is a need for a dedicated RA for the state of Perlis to clean up and verify the registry data.

11.5.2 Conclusion

The most common type of thalassaemia in Perlis is HbE-beta thalassaemia, followed by beta thalassaemia major and HbH disease. The median age group is 15-19.9 years old. DFX (87.84%) is the most common iron chelation therapy prescribed. Approximately 55% of patients on iron chelators achieve a serum ferritin level below 2500 ng/mL. A significant proportion of deaths were due to cardiac complication.

12 Pulau Pinang

12.1 Introduction

Penang has a population of 1,746,000 as of January 2018, with 480 thalassaemia patients. Hospital Pulau Pinang (HPP) and Hospital Seberang Jaya (HSJ) are the two hospitals providing care for all thalassaemia patients in the state (250 patients and 230 patients, respectively).

12.2 Patient Demographics

Data analysed were taken from patients with status of either alive, lost to follow-up or cured by transplant, and excludes deceased patients. The number of patients in Pulau Pinang is 451. Four patients were cured by transplant and 25 patients were lost to follow-up. The main causes of death were unrelated to thalassaemia, such as infections, intracranial bleed, tumours and others.

Table 12.1: Distribution of Patients in Pulau Pinang by Centre

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
Hospital Pulau Pinang	250	52.08
Hospital Seberang Jaya	230	47.92
Total	480	100.00

Figure 12.1: Distribution of Patients in Pulau Pinang by Centre

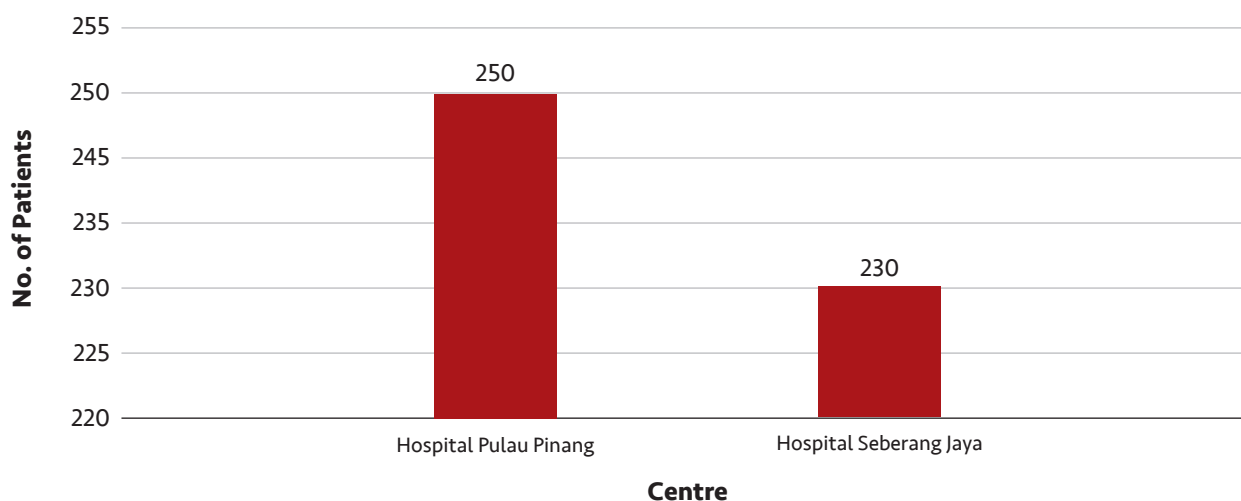


Table 12.2: Distribution of Patients in Pulau Pinang by Vital Status

Vital Status	Patients
Alive	451
Cumulative Reported Cured by Stem Cell Therapy (2008-2017)	4
Lost to Follow-up	25
Total	480
Deaths in 2017	1
Cumulative Reported Deaths	12

12 Pulau Pinang

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

Cause of Death	No. of Patients
Infections	6
Cardiac causes	1
Intracranial bleed	1
Tumours	1
MVA	2
Endocrine Complications	1
Unknown	0
Total	12

12.2.1 Age

The youngest patient in Pulau Pinang is 7 months old and the eldest is 85 years old. A total of 228 patients (47.50%) are below 20 years old. There are 21 new thalassaemia births between 2014-2018, although with reducing trend.

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

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	21	4.38
5 - 9.9	55	11.46
10 - 14.9	74	15.42
15 - 19.9	78	16.25
20 - 24.9	68	14.17
25 - 29.9	51	10.63
30 - 34.9	35	7.29
35 - 39.9	30	6.25
40 - 44.9	24	5.00
45 - 49.9	15	3.13
50 - 54.9	13	2.71
55 - 59.9	6	1.25
60 - 64.9	1	0.21
Above 65	9	1.88
Total	480	100.0

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

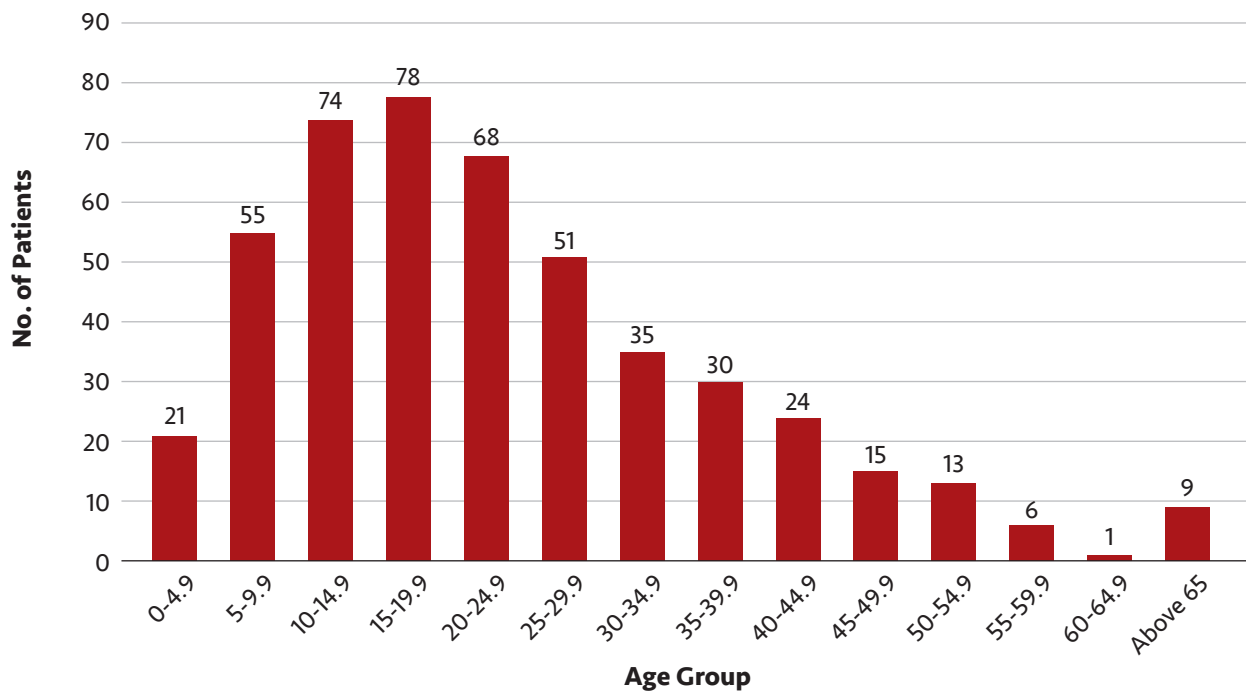


Table 12.5: Distribution of Patients in Pulau Pinang According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	21	Beta Thalassaemia Major	2	0.42
		Beta Thalassaemia Intermedia	3	0.63
		HbE-Beta Thalassaemia	9	1.88
		HbH Disease	5	1.04
		Others	2	0.42
5 - 9.9	55	Beta Thalassaemia Major	11	2.29
		Beta Thalassaemia Intermedia	3	0.63
		HbE-Beta Thalassaemia	24	5.00
		HbH Disease	13	2.71
		Others	4	0.83
10 -14.9	74	Beta Thalassaemia Major	20	4.17
		Beta Thalassaemia Intermedia	5	1.04
		HbE-Beta Thalassaemia	33	6.88
		HbH Disease	13	2.71
		Others	3	0.63
15 -19.9	78	Beta Thalassaemia Major	17	3.54
		Beta Thalassaemia Intermedia	2	0.42
		HbE-Beta Thalassaemia	39	8.13
		HbH Disease	16	3.33
		Others	4	0.83

12 Pulau Pinang

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
20 - 24.9	68	Beta Thalassaemia Major	19	3.96
		Beta Thalassaemia Intermedia	3	0.63
		HbE-Beta Thalassaemia	5	1.04
		HbH Disease	20	4.17
		Others	21	4.38
25 - 29.9	51	Beta Thalassaemia Major	17	3.54
		Beta Thalassaemia Intermedia	4	0.83
		HbE-Beta Thalassaemia	21	4.38
		HbH Disease	8	1.67
		Others	1	0.21
30 - 34.9	35	Beta Thalassaemia Major	7	1.46
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	17	3.54
		HbH Disease	6	1.25
		Others	4	0.83
35 - 39.9	30	Beta Thalassaemia Major	5	1.04
		Beta Thalassaemia Intermedia	7	1.46
		HbE-Beta Thalassaemia	10	2.08
		HbH Disease	7	1.46
		Others	1	0.21
40 - 44.9	24	Beta Thalassaemia Major	2	0.42
		Beta Thalassaemia Intermedia	2	0.42
		HbE-Beta Thalassaemia	11	2.29
		HbH Disease	7	1.46
		Others	2	0.42
45 - 49.9	15	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.21
		HbE-Beta Thalassaemia	3	0.63
		HbH Disease	11	2.29
		Others	0	0.00
50 - 54.9	13	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	0.42
		HbE-Beta Thalassaemia	5	1.04
		HbH Disease	6	1.25
		Others	0	0.00

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
55 - 59.9	6	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	3	0.63
		HbH Disease	3	0.63
		Others	0	0.00
60 - 64.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.21
		Others	0	0.00
Above 65	9	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	3	0.63
		HbH Disease	5	1.04
		Others	1	0.21
Total			480	100.0

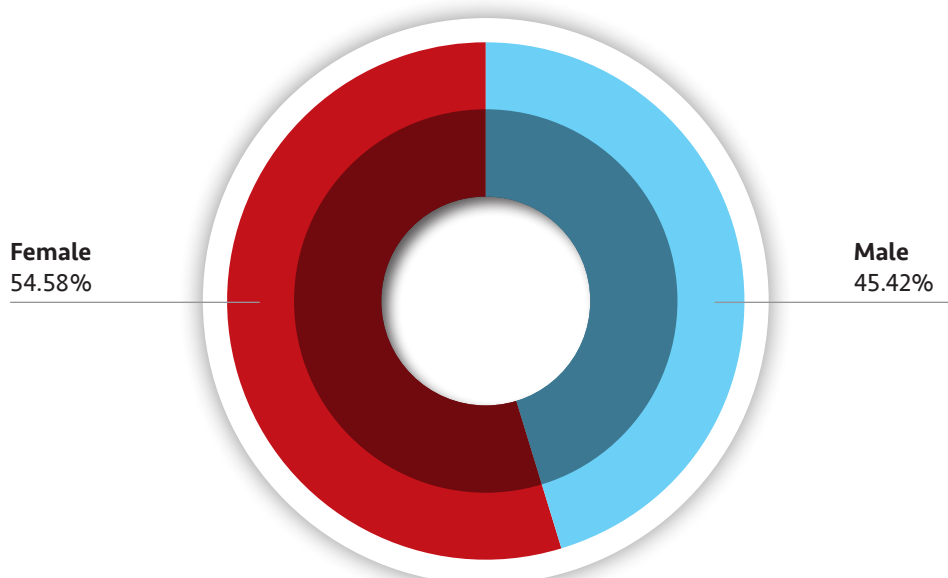
12.2.2 Gender

There are 218 male patients (45.42%) and 262 female patients (54.58%) in Pulau Pinang.

Table 12.6: Distribution of Patients in Pulau Pinang by Gender

Centre	Male		Female	
	No.	%	No.	%
HPP, Pulau Pinang	113	23.54	137	28.54
HSJ, Seberang Jaya	105	21.88	125	26.04
Total	218	45.42	262	54.58

Figure 12.3: Distribution of Patients in Pulau Pinang by Gender



12 Pulau Pinang

12.2.3 Ethnic Group

The Malays form the largest group of thalassaemia patients in HPP and HSJ with 379 patients (78.96%), followed by Chinese patients (84 patients, 17.50%), Indian patients (4 patients, 0.83%), Kadazan-Dusun (3 patients, 0.63%). The remaining 10 patients (2.08%) include Iban, Foreigner and Thai.

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

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	379	78.96
Chinese	84	17.50
Indian	4	0.83
Kadazan-Dusun	3	0.63
Others	10	2.08
Total	480	100.0

194

Figure 12.4: Distribution of Patients in Pulau Pinang by Ethnic Group

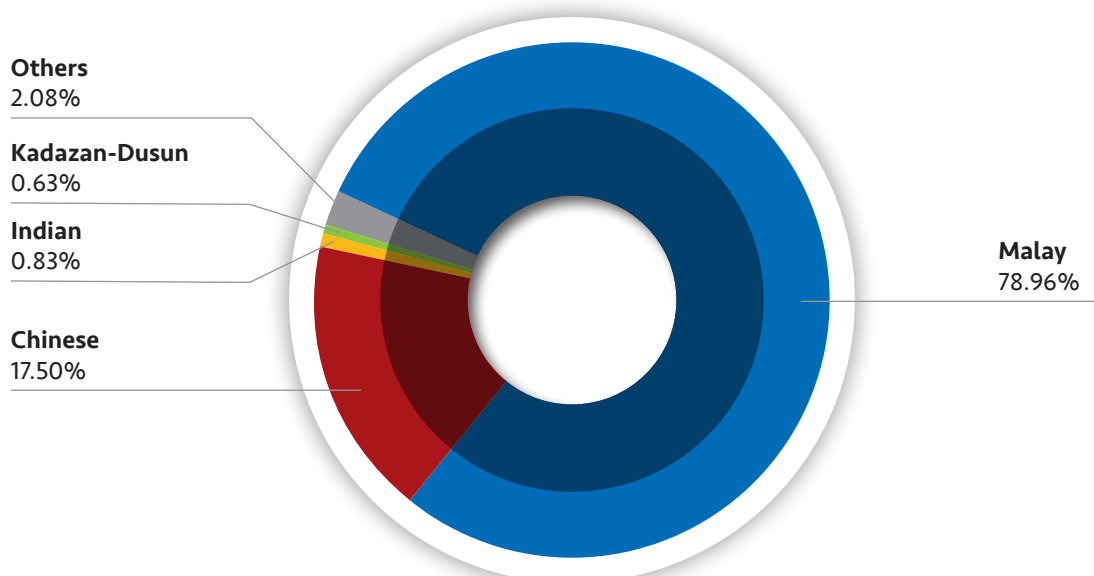


Figure 12.5: Distribution of Population of Pulau Pinang by Ethnic Group

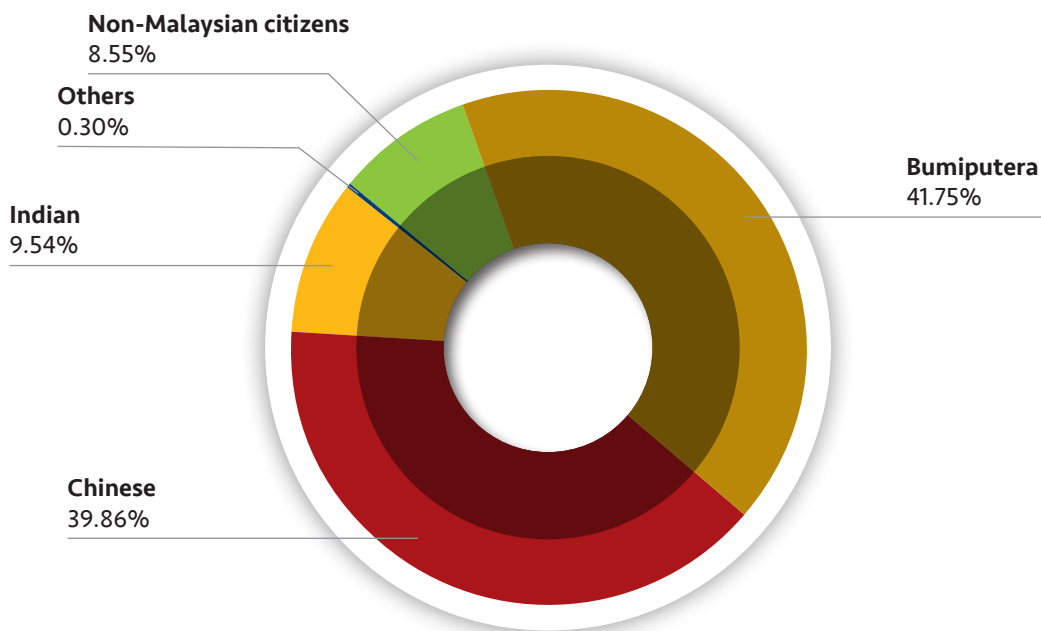


Table 12.8: Distribution of Patients in Pulau Pinang According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Kadazan		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Pulau Pinang	172	35.83	68	14.17	1	0.21	3	0.63	6	1.25
Hospital Seberang Jaya	207	43.13	16	3.33	3	0.63	0	0.00	4	0.83
Total	379	78.96	84	17.50	4	0.83	3	0.63	10	2.08

12.3 Diagnosis

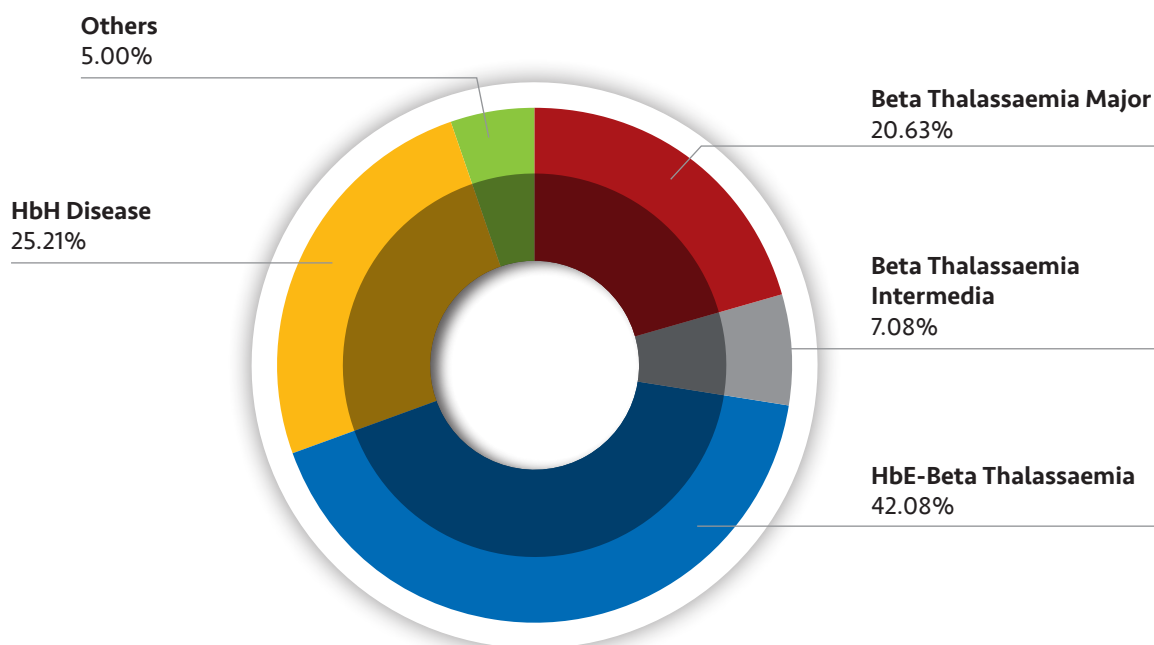
The most common diagnosis among patients in Pulau Pinang is HbE-beta thalassaemia with 202 patients (42.08%), followed by HbH disease with 121 patients (25.21%), beta thalassaemia major with 99 patients (20.63%), beta thalassaemia intermedia with 34 patients (7.08%). The remaining 24 patients (5.00%) consist of other diagnoses, as shown in Table 12.9.

Table 12.9: Distribution of Patients in Pulau Pinang by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	99	20.63
Beta Thalassaemia Intermedia	34	7.08
HbE-Beta Thalassaemia	202	42.08
HbH Disease	121	25.21
Others	24	5.00
Total	480	100.00

12 Pulau Pinang

Figure 12.6: Distribution of Patients in Pulau Pinang by Diagnosis



196

Table 12.10: Distribution of Patients in Pulau Pinang According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Pulau Pinang	52	10.83	18	3.75	97	20.21	72	15.00	11	2.29
Hospital Seberang Jaya	47	9.79	16	3.33	105	21.88	49	10.21	13	2.71
Total	99	20.63	34	7.08	202	42.08	121	25.21	24	5.00

Table 12.11: Distribution of Patients in Pulau Pinang According to Diagnosis by Ethnic Group

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	99	Malay	69	14.38
		Chinese	26	5.42
		Indian	1	0.21
		Kadazan-Dusun	2	0.42
		Others	1	0.21
Beta Thalassaemia Intermedia	34	Malay	28	5.83
		Chinese	5	1.04
		Indian	0	0.00
		Kadazan-Dusun	1	0.21
		Others	0	0.00

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
HbE-Beta Thalassaemia	202	Malay	182	37.92
		Chinese	14	2.92
		Indian	2	0.42
		Kadazan-Dusun	0	0.00
		Others	4	0.83
HbH Disease	121	Malay	86	17.92
		Chinese	31	6.46
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	4	0.83
Others	24	Malay	14	2.92
		Chinese	8	1.67
		Indian	1	0.21
		Kadazan-Dusun	0	0.00
		Others	1	0.21
Total			480	100

12.4 Treatment

12.4.1 Iron Chelation Therapy

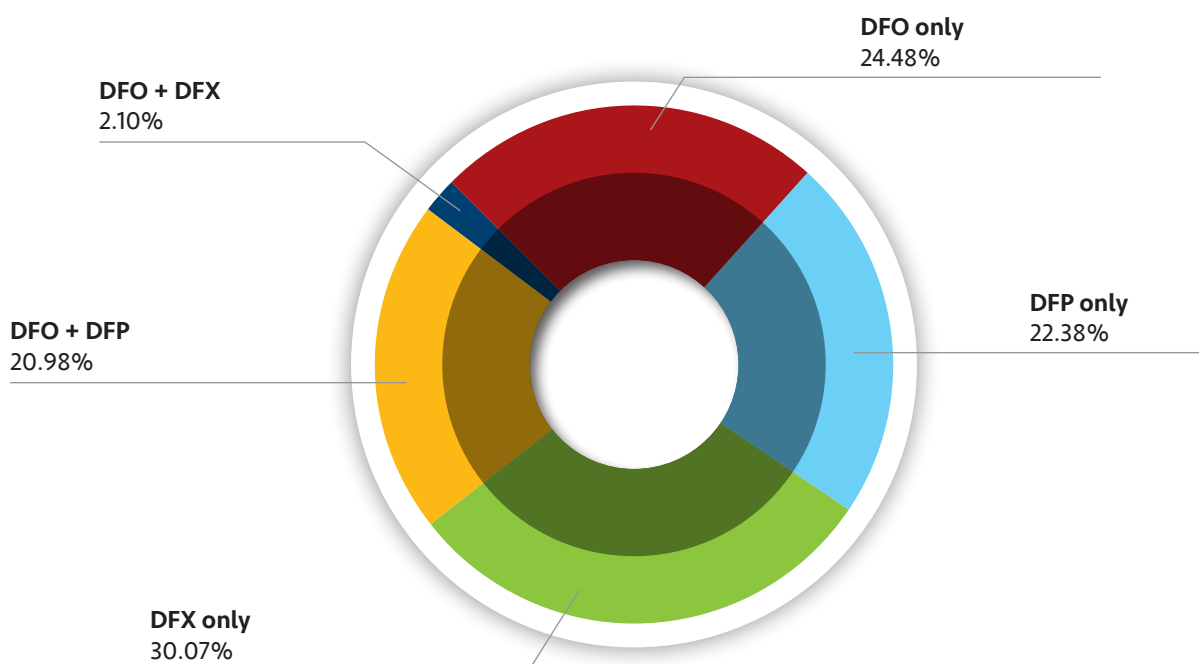
A total of 225 patients (46.88%) in Pulau Pinang have TDT, 211 patients (43.96%) are NTD and 44 patients (9.17%) have an unknown transfusion status. In addition, 286 patients (59.58%) received iron chelation therapy. Of this number, 216 patients (75.52%) have TDT and 66 patients (23.08%) have NTD. Seventy patients (24.48%) are on DFO, 64 patients (22.38%) are on DFP, 86 patients (30.07%) are on DFX, 60 patients (20.98%) are on a combination of DFO/DFP and 6 patients (2.10%) are on DFO/DFX.

Table 12.12: Distribution of Patients in Pulau Pinang According to Most Recent Serum Ferritin Level by Centre

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	70	24.48
DFP only	64	22.38
DFX only	86	30.07
DFO + DFP	60	20.98
DFP + DFX	0	0.00
DFO + DFX	6	2.10
DFO + DFP + DFX	0	0.00
Total	286	100.00

12 Pulau Pinang

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



198

Table 12.13: Distribution of Patients in Pulau Pinang According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Pulau Pinang	132	DFO only	19	6.64
		DFP only	31	10.84
		DFX only	40	13.99
		DFO + DFP	38	13.29
		DFP + DFX	0	0.00
		DFO + DFX	4	1.40
		DFO + DFP + DFX	0	0.00
Hospital Seberang Jaya	154	DFO only	51	17.83
		DFP only	33	11.54
		DFX only	46	16.08
		DFO + DFP	22	7.69
		DFP + DFX	0	0.00
		DFO + DFX	2	0.70
		DFO + DFP + DFX	0	0.00
Total			286	100.00

Table 12.14: Distribution of Patients in Pulau Pinang According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	5	DFO only	1	0.35
		DFP only	1	0.35
		DFX only	3	1.05
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5-9.9	30	DFO only	7	2.45
		DFP only	1	0.35
		DFX only	21	7.34
		DFO + DFP	1	0.35
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10-14.9	49	DFO only	16	5.59
		DFP only	1	0.35
		DFX only	28	9.79
		DFO + DFP	2	0.70
		DFP + DFX	0	0.00
		DFO + DFX	2	0.70
		DFO + DFP + DFX	0	0.00
15-19.9	49	DFO only	11	3.85
		DFP only	7	2.45
		DFX only	22	7.69
		DFO + DFP	9	3.15
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20-24.9	44	DFO only	7	2.45
		DFP only	9	3.15
		DFX only	6	2.10
		DFO + DFP	20	6.99
		DFP + DFX	0	0.00
		DFO + DFX	2	0.70
		DFO + DFP + DFX	0	0.00

12 Pulau Pinang

200

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
25-29.9	29	DFO only	8	2.80
		DFP only	9	3.15
		DFX only	1	0.35
		DFO + DFP	10	3.50
		DFP + DFX	0	0.00
		DFO + DFX	1	0.35
		DFO + DFP + DFX	0	0.00
30-34.9	20	DFO only	4	1.40
		DFP only	8	2.80
		DFX only	2	0.70
		DFO + DFP	6	2.10
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
35-39.9	18	DFO only	4	1.40
		DFP only	8	2.80
		DFX only	0	0.00
		DFO + DFP	6	2.10
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40-44.9	17	DFO only	3	1.05
		DFP only	11	3.85
		DFX only	1	0.35
		DFO + DFP	2	0.70
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-49.9	10	DFO only	4	1.40
		DFP only	3	1.05
		DFX only	0	0.00
		DFO + DFP	2	0.70
		DFP + DFX	0	0.00
		DFO + DFX	1	0.35
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
50-54.9	9	DFO only	2	0.70
		DFP only	5	1.75
		DFX only	1	0.35
		DFO + DFP	1	0.35
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	4	DFO only	1	0.35
		DFP only	1	0.35
		DFX only	1	0.35
		DFO + DFP	1	0.35
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60-64.9	1	DFO only	1	0.35
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	1	DFO only	1	0.35
		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			286	100.00

12 Pulau Pinang

12.4.2 Serum Ferritin Level

There are 135 regularly transfused patients in Pulau Pinang who had their serum ferritin levels measured in 2018. Twenty-six (19.26%) of the TDT patients had serum ferritin level below 1000 ng/mL, 70 patients (51.85%) have serum ferritin level below 2500 ng/mL, and 65 patients (48.15%) have a serum ferritin level above 2500 ng/mL.

Table 12.15: Distribution of Patients in Pulau Pinang According to Most Recent Serum Ferritin Level by Centre

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Pulau Pinang	98	22	16.30	35	25.93	19	14.07	20	14.81	2	1.48
Hospital Seberang Jaya	37	4	2.96	9	6.67	19	14.07	5	3.70	0	0.00
Total	135	26	19.26	44	32.59	38	28.15	25	18.52	2	1.48

12.5 Observation and Comments

1. Nearly half (47.50%) of the thalassaemia patients in Pulau Pinang are under 20 years old and 16.25% are in the transitional age group (15-19.9 years old).
2. The ethnic distribution of thalassaemia patients in Pulau Pinang is mainly Malay (78.96%), followed by Chinese (17.50%), Indian (0.83%), in contrast to the ethnic distribution of population in Pulau Pinang, that is 42% Bumiputera, 40% Chinese, and 9.6% Indian. This indicates a higher prevalence rate of thalassaemia among the Malays in Pulau Pinang.
3. HbE-beta thalassaemia is the most common type of diagnosis (42.08%) and contributes to a significant number of TDT patients, who require chelation therapy.
4. The most common cause of death among thalassaemia patients in Pulau Pinang is infection, afflicting 6 out of 12 deceased patients.
5. Most patients are well-chelated, with 19.26% of the TDT patients achieving serum ferritin level below 1000 ng/mL and 51.85% achieving serum ferritin level below 2500 ng/mL.

12.5.1 Recommendations

1. The number of new thalassaemia births remained static in the past few years. Therefore, cooperation with non-governmental organisations to increase awareness and acceptance of antenatal screening need to be enhanced.
2. As HbE thalassaemia is the most prevalent subtype of thalassaemia, further studies on genotype-phenotype correlation and disease modifying factors is needed.
3. Infection remains the main cause of death of thalassaemia patients. Therefore, education of the patients and healthcare workers should be emphasised to ensure early and adequate treatment of infections in thalassaemia patients.
4. Budget planning should take into consideration that thalassaemia patients are expected to have improved survival into adulthood and budget distribution should take into account the group of transitional patients from paediatrics to adult healthcare.
5. Additional budget is needed to ensure availability of all iron chelators and provision of sufficient infusion pumps.
6. MTAC services are needed as it may help to improve adherence in poorly chelated patients.
7. Aggregate data for the whole state should be made accessible for the paediatricians and haematologists in charge of thalassaemia care to help identify areas for improvement and research.
8. Data on serum ferritin level, diagnosis and genotype were incomplete and need to be updated.

13 Sabah

13.1 Introduction

Sabah is the second largest state in Malaysia after Sarawak, with a land area of 73,619 km². According to the Department of Statistics Malaysia's population census of year 2010, the population of Sabah was 3,206,742.

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

Table 13.1: Population of Sabah by Ethnicity

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

* "Malay" consists of Bruneian Malays, Kedayan, Banjar, Cocos and Peninsular Malays.

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

Adapted from Department of Statistics Malaysia. 2010 Population and Housing Census of Malaysia.

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

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

Division Name	Districts	Area (km ²)	Population (2010)	No. of Thalassemia Patients	Prevalence of Thalassemia (Per 100,000 Population)	
1	Pantai Barat (Kota Kinabalu)	Kota Belud, Kota Kinabalu, Papar, Penampang, Putatan, Ranau, Tuaran	7,588	1,067,589	874	81.87
2	Pedalaman (Keningau)	Beaufort, Keningau, Nabawan, Kuala Penyu, Sipitang, Tambunan, Tenom	18,298	424,534	312	73.49
3	Kudat	Kota Marudu, Kudat, Pitas	4,623	192,457	286	148.60
4	Sandakan	Beluran, Kinabatangan, Sandakan, Tongod	28,205	702,207	239	34.02
5	Tawau	Kunak, Lahad Datu, Semporna, Tawau	14,905	819,955	103	12.56
Total		-	3,206,742	1814	-	

13.2 Patient Demographics

Total number of thalassaemia patients in Sabah registered in the MTR until November 2018 is 1814.

Table 13.3: Distribution of Thalassaemia Patients in Each Treatment Centre in Sabah and Estimated Prevalence Rate in the Respective City/Town

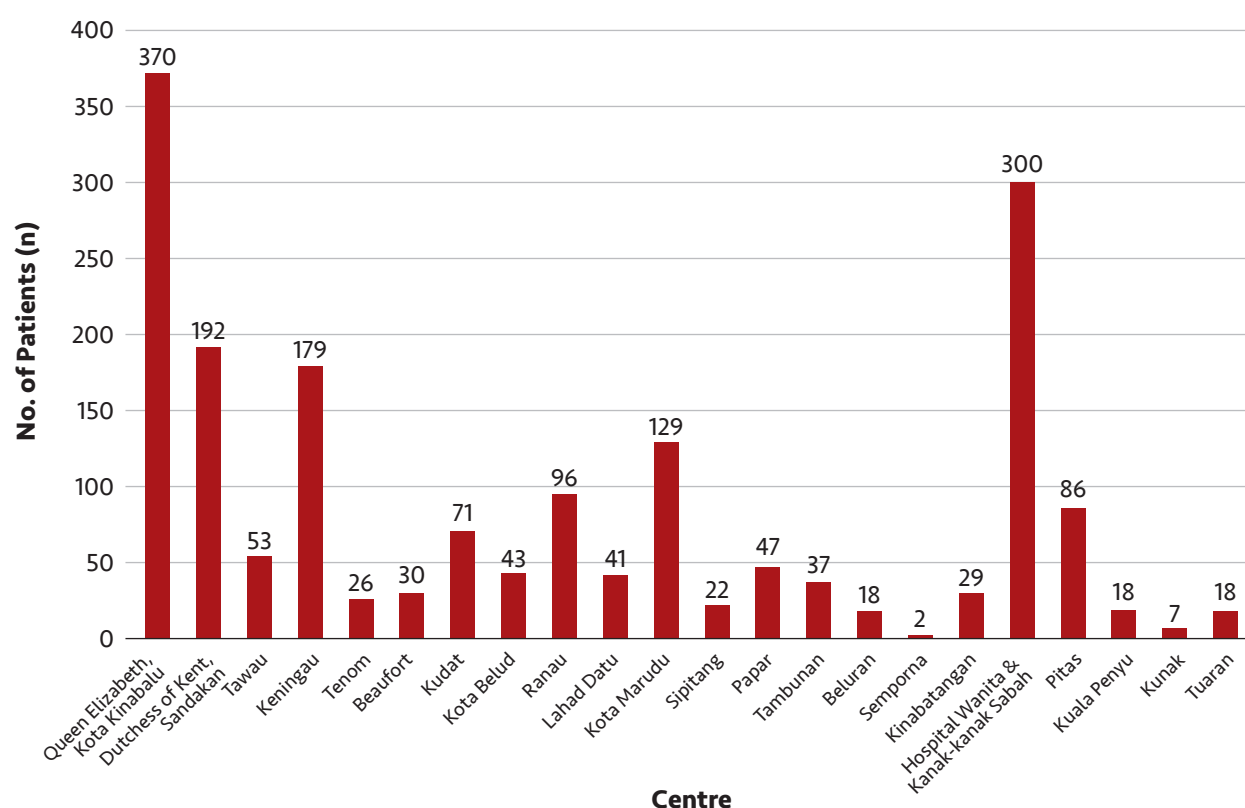
Centre	Total No. of Patients	Percentage (%)	Population (2010)	Prevalence rate (Per 100,000 population)
Hospital Wanita & Kanak-Kanak Sabah, Likas	300	16.54	750,175	39.99
Queen Elizabeth, Kota Kinabalu	370	20.40		
Tuaran	18	0.99		
Duchess of Kent, Sandakan	192	10.58	409,056	46.94
Keningau	179	9.87	210,044	85.22
Kota Marudu	129	7.11	68,289	188.90
Ranau	96	5.29	95,800	100.21
Kudat	71	3.91	85,404	83.13
Pitas	86	4.74	38,764	221.86
Kota Belud	43	2.37	93,180	46.15
Papar	47	2.59	128,434	36.59
Tawau	53	2.92	412,375	12.85
Tenom	26	1.43	56,597	45.94
Tambunan	37	2.04	36,297	101.94
Lahad Datu	41	2.26	206,861	19.82
Kinabatangan*	29	1.60	186,519	15.55
Beaufort	30	1.65	66,406	45.18
Beluran	18	0.99	106,632	16.88
Kuala Penyu	18	0.99	19,426	92.66
Sipitang	22	1.21	35,764	61.51
Kunak	7	0.39	62,851	11.14
Semporna	2	0.11	137,868	1.45
Total	1814	100.00	3,206,742	

* Total Population of Kinabatangan and Tongod

The three hospitals with the highest number of patients in Sabah are Hospital Wanita & Kanak-Kanak Sabah (HWKKS), Kota Kinabalu with 300 patients (16.54%), Hospital Queen Elizabeth (HQE) with 370 patients (20.40%) and Hospital Duchess of Kent (HDOK), Sandakan with 192 patients (10.58%).

The pattern of distribution of thalassaemia patients by ethnic group follows closely the distribution of the ethnic groups of Sabah, which have a high carrier rate of beta thalassaemia, i.e. the 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 bigger hospitals in the Southeast of Sabah, e.g. Tawau and Lahad Datu Hospitals have relatively fewer patients. This phenomenon is due to the relatively low density of Kadazan-Dusun, Rungus, Bajau and Murut populations in these towns.

Figure 13.1: Distribution of Patients in Sabah by Centre



205

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Table 13.4: Distribution of Patients in Sabah by Vital Status

Vital Status	Patients
Alive	1703
Cumulative Reported Cured by Stem Cell Therapy (2008-2018)	36
Lost to Follow-up	75
Total	1814
Deaths in 2018	18
Cumulative Reported Deaths	303

Table 13.5: Cumulative Causes of Death Since 2007 in Sabah

Cause of Death	No. of Patients
Cardiac Cause	98
Endocrine Complication	6
Infection	64
Others	44
Unknown*	91
Total	303

*Missing data.

There were only 18 deaths reported in 2018, which is a slight decrease from 21 deaths reported in 2017.

13.2.1 Age

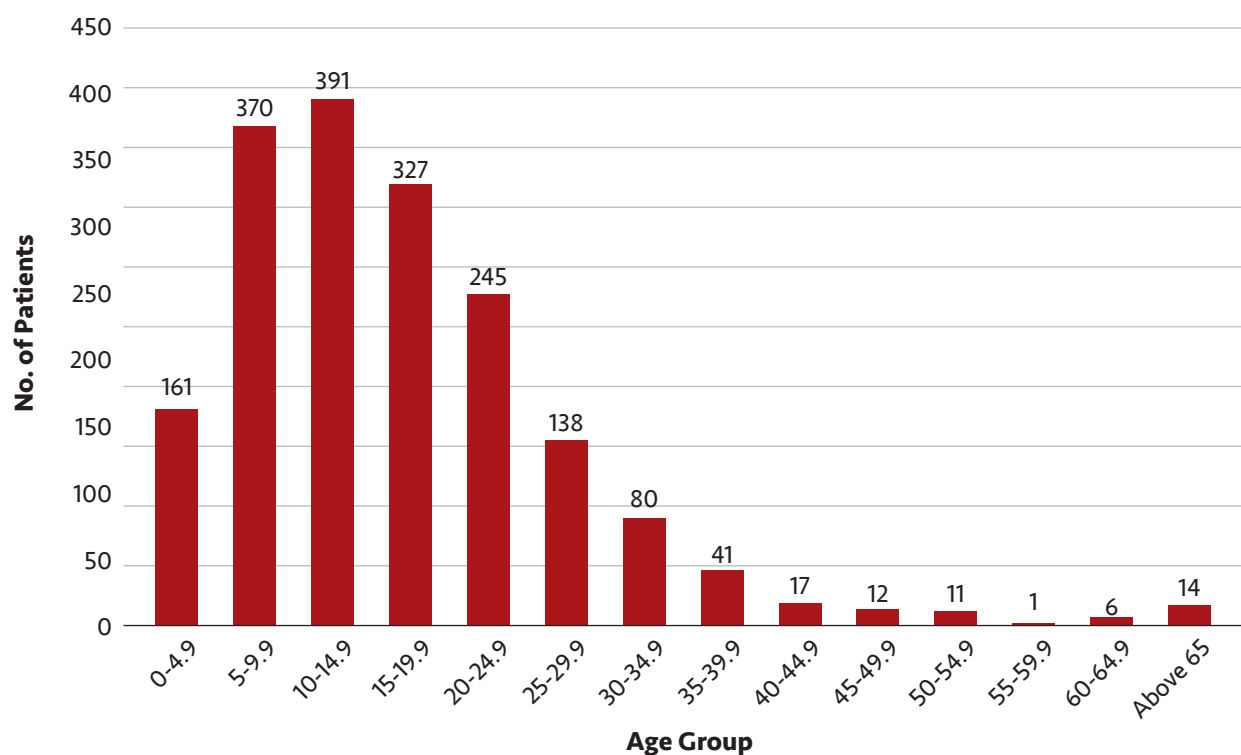
In current 2018 statistics, the age group with the highest number of thalassaemia patients is 10-14.9 years. In 2017, the age group with highest number of thalassaemia patients was 6-10 years. This reflects an improvement in patients' survival rates. The oldest surviving thalassaemia major patient in Sabah is currently 40 years old.

In 2018, the number of thalassaemia patients younger than 15 years old continue to increase in number, compared to in 2017. However, the overall percentage of patients younger than 15 years old in 2015 had reduced in view of increasing survival rates of whole cohorts of thalassaemia patients in 2018.

Table 13.6: Distribution of Patients in Sabah by Age Group

Age Group (years)	Patient Distribution				Percentage (%)
	No. of Patients with Beta Thalassaemia	No. of Patients with Alpha Thalassaemia	Others	Total	
0 - 4.9	155	4	2	161	8.88
5 - 9.9	354	15	1	370	20.40
10 -14.9	380	10	1	391	21.55
15 -19.9	318	8	1	327	18.03
20 - 24.9	240	5	0	245	13.51
25 - 29.9	131	7	0	138	7.61
30 - 34.9	67	12	1	80	4.41
35 - 39.9	34	7	0	41	2.26
40 - 44.9	15	2	0	17	0.94
45 - 49.9	7	5	0	12	0.66
50 - 54.9	9	2	0	11	0.61
55 - 59.9	1	0	0	1	0.06
60 - 64.9	1	5	0	6	0.33
Above 65	5	9	0	14	0.77
Total	1717	91	6	1814	100.00

Figure 13.2: Distribution of Patients in Sabah by Age Group



207

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Table 13.7: Distribution of Patients in Sabah According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	161	Beta Thalassaemia Major	132	7.28
		Beta Thalassaemia Intermedia	16	0.88
		HbE-Beta Thalassaemia	7	0.39
		HbH Disease	4	0.22
		Others	2	0.11
5 - 9.9	370	Beta Thalassaemia Major	286	15.77
		Beta Thalassaemia Intermedia	48	2.65
		HbE-Beta Thalassaemia	20	1.10
		HbH Disease	15	0.83
		Others	1	0.06
10 -14.9	391	Beta Thalassaemia Major	298	16.43
		Beta Thalassaemia Intermedia	57	3.14
		HbE-Beta Thalassaemia	25	1.38
		HbH Disease	10	0.55
		Others	1	0.06
15 -19.9	327	Beta Thalassaemia Major	261	14.39
		Beta Thalassaemia Intermedia	45	2.48
		HbE-Beta Thalassaemia	12	0.66
		HbH Disease	8	0.44
		Others	1	0.06

13 Sabah

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
20 - 24.9	245	Beta Thalassaemia Major	202	11.14
		Beta Thalassaemia Intermedia	34	1.87
		HbE-Beta Thalassaemia	4	0.22
		HbH Disease	5	0.28
		Others	0	0.00
25 - 29.9	138	Beta Thalassaemia Major	91	5.02
		Beta Thalassaemia Intermedia	27	1.49
		HbE-Beta Thalassaemia	13	0.72
		HbH Disease	7	0.39
		Others	0	0.00
30 - 34.9	80	Beta Thalassaemia Major	39	2.15
		Beta Thalassaemia Intermedia	18	0.99
		HbE-Beta Thalassaemia	10	0.55
		HbH Disease	12	0.66
		Others	1	0.06
35 - 39.9	41	Beta Thalassaemia Major	12	0.66
		Beta Thalassaemia Intermedia	16	0.88
		HbE-Beta Thalassaemia	6	0.33
		HbH Disease	7	0.39
		Others	0	0.00
40 - 44.9	17	Beta Thalassaemia Major	4	0.22
		Beta Thalassaemia Intermedia	6	0.33
		HbE-Beta Thalassaemia	5	0.28
		HbH Disease	2	0.11
		Others	0	0.00
45 - 49.9	12	Beta Thalassaemia Major	1	0.06
		Beta Thalassaemia Intermedia	4	0.22
		HbE-Beta Thalassaemia	2	0.11
		HbH Disease	5	0.28
		Others	0	0.00
50 - 54.9	11	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	0.11
		HbE-Beta Thalassaemia	7	0.39
		HbH Disease	2	0.11
		Others	0	0.00
55 - 59.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	1	0.06
		HbH Disease	0	0.00
		Others	0	0.00

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
60 - 64.9	6	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	1	0.06
		HbH Disease	5	0.28
		Others	0	0.00
Above 65	14	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	3	0.17
		HbE-Beta Thalassaemia	2	0.11
		HbH Disease	9	0.50
		Others	0	0.00
Total			1814	100.00

Beta thalassaemia major constitutes the majority diagnosis in Sabah. Up to mid-November 2018, 73.10% of total number of patients was diagnosed with beta thalassaemia major, and 15.21% of patients were diagnosed with beta thalassaemia intermedia. HbE-beta thalassaemia constitutes much fewer cases in Sabah compared to West Malaysia. This observation is consistent with the discovery of the predominant affected severe form of mutation—a 45-kb beta globin gene deletion (Filipino deletion), occurring in homozygous form in more than 90% of Sabahan Kadazan-Dusuns with beta thalassaemia syndrome.

In comparison with MTR data reported in year 2017, there was an increase of 22 thalassaemia patients in year 2018, including thalassaemia major patients. This increased number of patients impose increased workload and healthcare burden to the treatment centres in Sabah.

13.2.2 Gender

The gender distribution of thalassaemia patients in Sabah is near equal, i.e. 939 male patients (51.76%) and 875 female patients (48.24%), as is consistent with thalassaemia syndrome as an autosomal recessive disease.

Figure 13.3: Distribution of Patients in Sabah by Gender

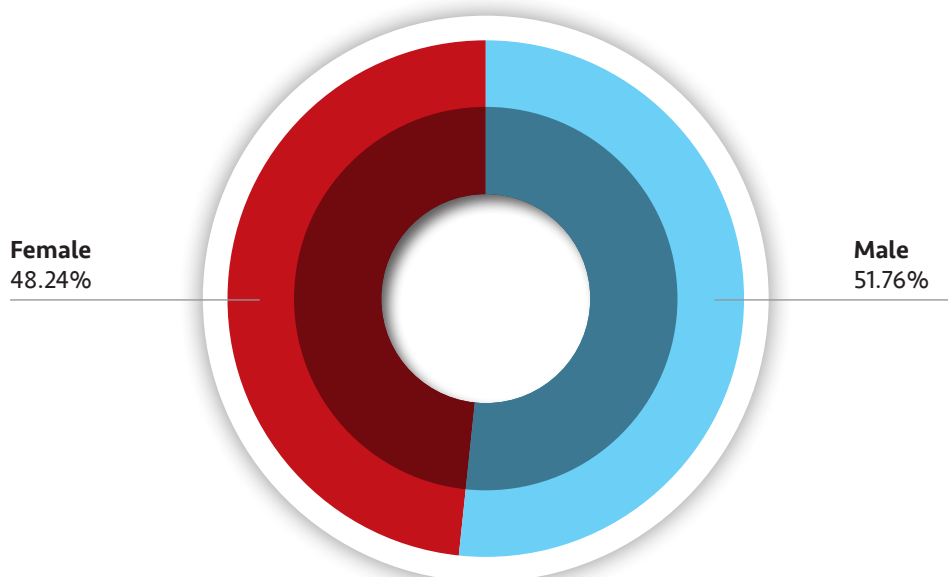


Table 13.8: Distribution of Patients in Sabah by Gender

Centre	Total	Male		Female	
		No.	%	No.	%
Queen Elizabeth, Kota Kinabalu	370	169	9.32	201	11.08
Duchess of Kent, Sandakan	192	93	5.13	99	5.46
Tawau	53	24	1.32	29	1.60
Keningau	179	94	5.18	85	4.69
Tenom	26	11	0.61	15	0.83
Beaufort	30	18	0.99	12	0.66
Kudat	71	39	2.15	32	1.76
Kota Belud	43	26	1.43	17	0.94
Ranau	96	56	3.09	40	2.21
Lahad Datu	41	13	0.72	28	1.54
Kota Marudu	129	65	3.58	64	3.53
Sipitang	22	14	0.77	8	0.44
Papar	47	34	1.87	13	0.72
Tambunan	37	22	1.21	15	0.83
Beluran	18	12	0.66	6	0.33
Semporna	2	0	0.00	2	0.11
Kinabatangan	29	15	0.83	14	0.77
Hospital Wanita & Kanak-Kanak Sabah, Likas	300	159	8.77	141	7.77
Pitas	86	43	2.37	43	2.37
Kuala Penyu	18	13	0.72	5	0.28
Kunak	7	5	0.28	2	0.11
Tuaran	18	14	0.77	4	0.22
Total	1814	939	51.76	875	48.24

13.2.3 Ethnic Group

The Kadazan-Dusun form the largest group of patients in Sabah with 858 patients (47.30%), followed by Pribumi Sabah with 202 patients (11.14%).

Table 13.9: Distribution of Patients in Sabah by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	101	5.57
Chinese	96	5.29
Indian	1	0.06
Kadazan-Dusun	858	47.30
Others:		
Rungus	111	6.12
Bajau	185	10.20
Murut	139	7.66
Mixed	28	1.54
Others	33	1.82
Sino-Kadazan	27	1.49
Foreigner	26	1.43
Iban	4	0.22
Bidayuh	1	0.06
Thai	1	0.06
Pribumi Sabah	202	11.14
Pribumi Sarawak	1	0.06
Unknown	0	0.00
Total	1814	100.00

Figure 13.4: Distribution of Patients in Sabah by Ethnic Group

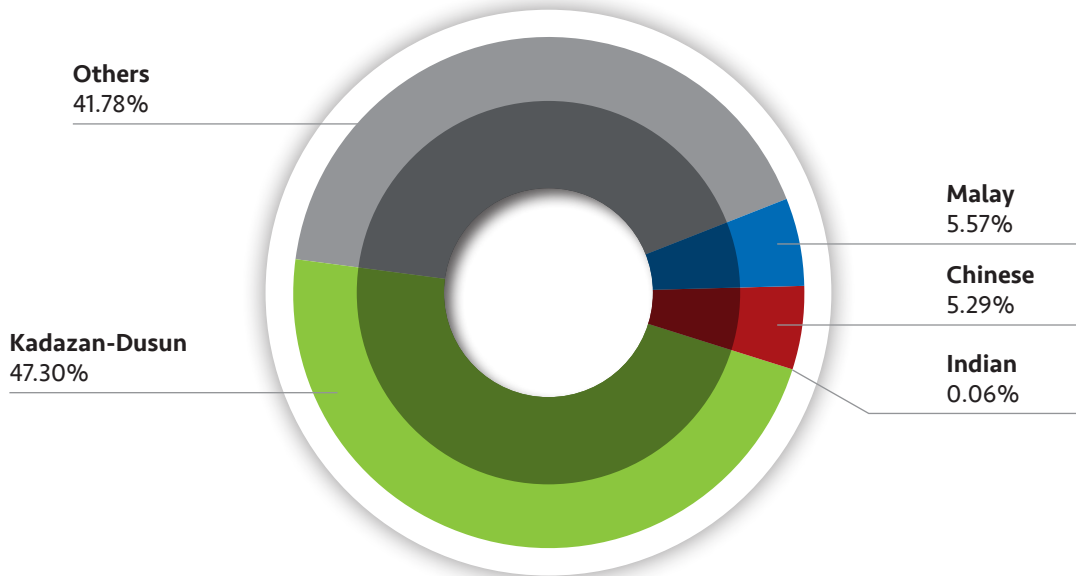


Figure 13.5: Distribution of Patients in Sabah by Ethnic Sub-Group

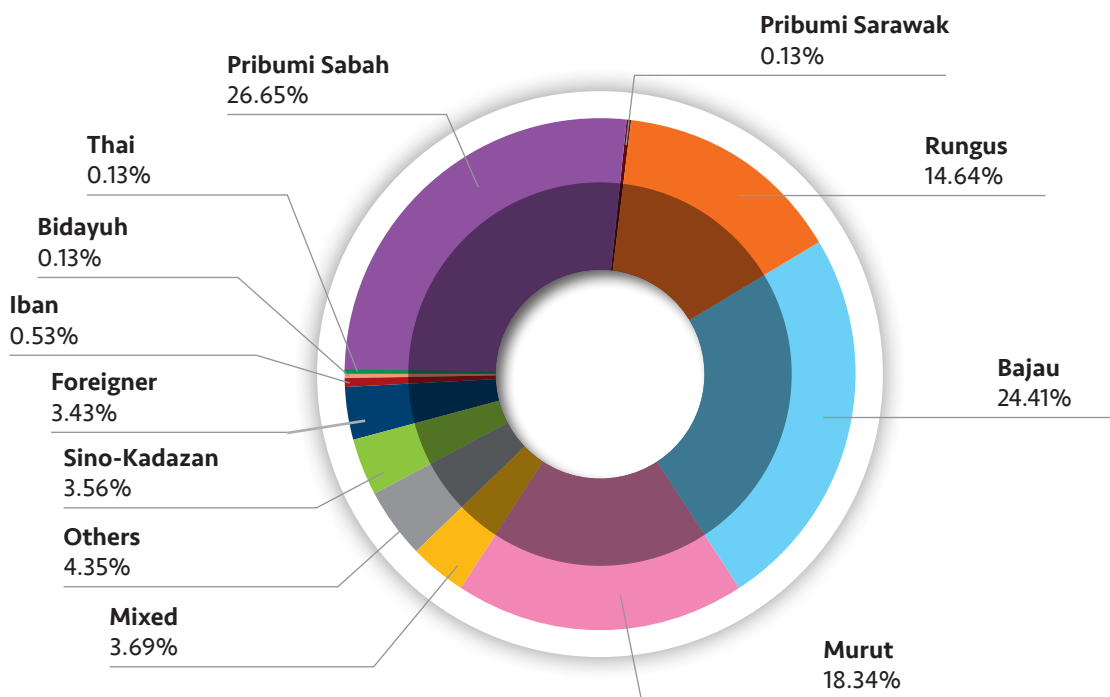


Table 13.10: Distribution of Patients in Sabah According to Ethnic Group by Centre

Centre	Kadazan-Dusun		Rungus		Malay		Bajau		Murut		Chinese		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
HQE	162	8.93	27	1.49	27	1.49	56	3.09	11	0.61	49	2.70	38	2.09
DOK	56	3.09	0	0.00	17	0.94	23	1.27	0	0.00	7	0.39	89	4.91
Tawau	8	0.44	0	0.00	6	0.33	11	0.61	0	0.00	7	0.39	21	1.16
Keningau	87	4.80	0	0.00	1	0.06	1	0.06	82	4.52	4	0.22	4	0.22
Tenom	2	0.11	0	0.00	2	0.11	2	0.11	17	0.94	2	0.11	1	0.06
Beaufort	6	0.33	0	0.00	4	0.22	2	0.11	2	0.11	0	0.00	16	0.88
Kudat	8	0.44	42	2.32	0	0.00	7	0.39	0	0.00	4	0.22	10	0.55
Kota Belud	35	1.93	0	0.00	1	0.06	4	0.22	1	0.06	0	0.00	2	0.11
Ranau	96	5.29	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Lahad Datu	5	0.28	0	0.00	4	0.22	15	0.83	0	0.00	1	0.06	16	0.88
Kota Marudu	96	5.29	11	0.61	1	0.06	5	0.28	0	0.00	1	0.06	15	0.83
Sipitang	0	0.00	0	0.00	3	0.17	1	0.06	14	0.77	0	0.00	4	0.22
Papar	23	1.27	3	0.17	7	0.39	5	0.28	3	0.17	0	0.00	6	0.33
Tambunan	33	1.82	0	0.00	0	0.00	0	0.00	2	0.11	0	0.00	2	0.11
Beluran	14	0.77	0	0.00	0	0.00	1	0.06	0	0.00	0	0.00	3	0.17
Semporna	0	0.00	0	0.00	0	0.00	1	0.06	0	0.00	0	0.00	1	0.06
Kinabatangan	1	0.06	0	0.00	3	0.17	5	0.28	0	0.00	0	0.00	20	1.10
HWKKL	160	8.82	12	0.66	22	1.21	35	1.93	7	0.39	19	1.05	45	2.48
Pitas	39	2.15	16	0.88	2	0.11	2	0.11	0	0.00	1	0.06	26	1.43
Kuala Penyu	11	0.61	0	0.00	1	0.06	3	0.17	0	0.00	0	0.00	3	0.17
Kunak	0	0.00	0	0.00	0	0.00	5	0.28	0	0.00	0	0.00	2	0.11
Tuaran	16	0.88	0	0.00	0	0.00	1	0.06	0	0.00	1	0.06	0	0.00
Total	858	47.30	111	6.12	101	5.57	185	10.20	139	7.66	96	5.29	324	17.86

A significant percentage of thalassaemia patients in Sabah are of Kadazan-Dusun descent (47.3% of total number of patients). This is disproportionately high compared to the percentage of Kadazan-Dusuns in the population of Sabah. There are also disproportionately high prevalence of thalassaemia cases amongst the ethnicities of Rungus (6.12%) and Murut (7.66%).

The predominance of thalassaemia syndrome amongst the ethnic Kadazan-Dusun and in the West Coast of Sabah have implications on allocation of resources towards effective thalassaemia control.

13.3 Diagnosis

Beta thalassaemia major constitutes the majority diagnosis in Sabah. Up to mid-November 2018, 73.15% of thalassaemia patients in Sabah were diagnosed with beta thalassaemia major, and 15.21% patients were diagnosed with beta thalassaemia intermedia. HbE-beta thalassaemia constitutes much fewer cases in Sabah as compared to West Malaysia. This observation is consistent with the discovery of the predominant affected severe form of mutation—a 45-kb beta globin gene deletion (Filipino deletion), occurring in homozygous form in more than 90% of Sabahan Kadazan-Dusuns with beta thalassaemia syndrome.

Table 13.11: Distribution of Patients in Sabah by Diagnosis

Diagnosis	No. of Patients	Percentage (%)
Beta Thalassaemia Major	1327	73.15
Beta Thalassaemia Intermedia	276	15.21
HbE-Beta Thalassaemia	115	6.34
HbH Disease	91	5.02
Others	5	0.28
Total	1814	100.00

Figure 13.6: Distribution of Patients in Sabah by Diagnosis

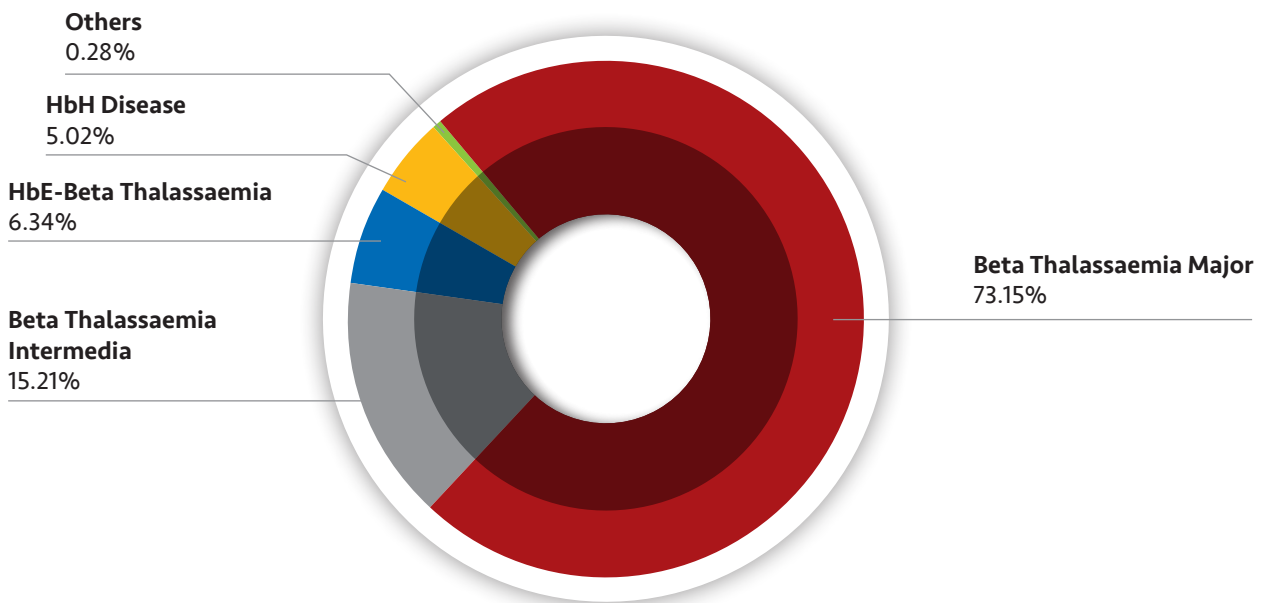


Table 13.12: Distribution of Patients in Sabah According to Diagnosis by Centre

Diagnosis	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Beaufort	23	1.27	7	0.39	0	0.00	0	0.00	0	0.00
Beluran	18	0.99	0	0.00	0	0.00	0	0.00	0	0.00
DOK	115	6.34	46	2.54	22	1.21	9	0.50	0	0.00
HWKKS	210	11.58	62	3.42	15	0.83	12	0.66	1	0.06
Keningau	165	9.10	9	0.50	3	0.17	1	0.06	1	0.06
Kinabatangan	24	1.32	4	0.22	1	0.06	0	0.00	0	0.00
Kota Belud	35	1.93	3	0.17	5	0.28	0	0.00	0	0.00
Kota Marudu	109	6.01	15	0.83	4	0.22	1	0.06	0	0.00
Kuala Penyu	11	0.61	6	0.33	0	0.00	0	0.00	1	0.06
Kudat	53	2.92	5	0.28	5	0.28	8	0.44	0	0.00
Kunak	2	0.11	5	0.28	0	0.00	0	0.00	0	0.00
Lahad Datu	23	1.27	10	0.55	5	0.28	2	0.11	1	0.06
Papar	38	2.09	7	0.39	2	0.11	0	0.00	0	0.00
Pitas	74	4.08	5	0.28	6	0.33	1	0.06	0	0.00
HQE	220	12.13	60	3.31	40	2.21	49	2.70	1	0.06
Ranau	87	4.80	7	0.39	2	0.11	0	0.00	0	0.00
Semporna	2	0.11	0	0.00	0	0.00	0	0.00	0	0.00
Sipitang	18	0.99	3	0.17	0	0.00	1	0.06	0	0.00
Tambunan	35	1.93	2	0.11	0	0.00	0	0.00	0	0.00
Tawau	26	1.43	17	0.94	4	0.22	6	0.33	0	0.00
Tenom	22	1.21	2	0.11	1	0.06	1	0.06	0	0.00
Tuaran	17	0.94	1	0.06	0	0.00	0	0.00	0	0.00
Total	1327	73.15	276	15.21	115	6.34	91	5.02	5	0.28

215

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Table 13.13: Distribution of Patients in Sabah According to Diagnosis by Ethnic Group

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	1327	Malay	55	3.03
		Chinese	47	2.59
		Indian	1	0.06
		Kadazan-Dusun	747	41.18
		Others:		
		Rungus	97	5.35
		Bajau	77	4.24
		Murut	124	6.84
		Mixed	14	0.77
		Others	165	9.10

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	276	Malay	22	1.21
		Chinese	8	0.44
		Indian	0	0.00
		Kadazan-Dusun	82	4.52
		Others:		
		Rungus	12	0.66
		Bajau	51	2.81
		Murut	12	0.66
		Mixed	10	0.55
		Others	79	4.36
HbE-Beta Thalassaemia	115	Malay	16	0.88
		Chinese	6	0.33
		Indian	0	0.00
		Kadazan-Dusun	21	1.16
		Others:		
		Rungus	1	0.06
		Bajau	35	1.93
		Murut	1	0.06
		Mixed	4	0.22
		Others	31	1.71
HbH Disease	91	Malay	7	0.39
		Chinese	34	1.87
		Indian	0	0.00
		Kadazan-Dusun	7	0.39
		Others:		
		Rungus	1	0.06
		Bajau	20	1.10
		Murut	2	0.11
		Mixed	0	0.00
		Others	20	1.10
Others	5	Malay	1	0.06
		Chinese	1	0.06
		Indian	0	0.00
		Kadazan-Dusun	1	0.06
		Others:		
		Rungus	0	0.00
		Bajau	2	0.11
		Murut	0	0.00
		Mixed	0	0.00
		Others	0	0.00
Total			1814	100.00

13.4 Treatment

13.4.1 Iron Chelation Therapy

The distribution patients in Sabah according to type of iron chelator received are as shown in Table 13.14.

Table 13.14: Distribution of Patients in Sabah by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	349	28.75
DFP only	142	11.70
DFX only	339	27.92
DFO + DFP	248	20.43
DFP + DFX	44	3.62
DFO + DFX	59	4.86
DFO + DFP + DFX	33	2.72
Total	1214	100.00

DFO is the most common type of iron chelator prescribed (28.75%), followed by DFX (27.92%) and a combination of DFO and DFP (20.43%). DFX is mainly prescribed to the paediatric patients. Many older thalassaemia major patients in Sabah started chelation therapy later in life, i.e. when the government provided free chelation treatment beginning in 2006. Many of these patients already had severe iron overload; hence these patients are treated with a combination of DFO and DFP. A total of 606 patients in Sabah was registered as 'Not on Iron Chelation Therapy'. This is likely to be false, caused by incomplete data entry or technical errors and need further verification.

Table 13.15: Distribution of Patients in Sabah According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Queen Elizabeth, Kota Kinabalu	238	DFO only	118	9.72
		DFP only	21	1.73
		DFX only	2	0.16
		DFO + DFP	75	6.18
		DFP + DFX	0	0.00
		DFO + DFX	5	0.41
		DFO + DFP + DFX	17	1.40
Duchess of Kent, Sandakan	151	DFO only	58	4.78
		DFP only	21	1.73
		DFX only	39	3.21
		DFO + DFP	22	1.81
		DFP + DFX	4	0.33
		DFO + DFX	6	0.49
		DFO + DFP + DFX	1	0.08

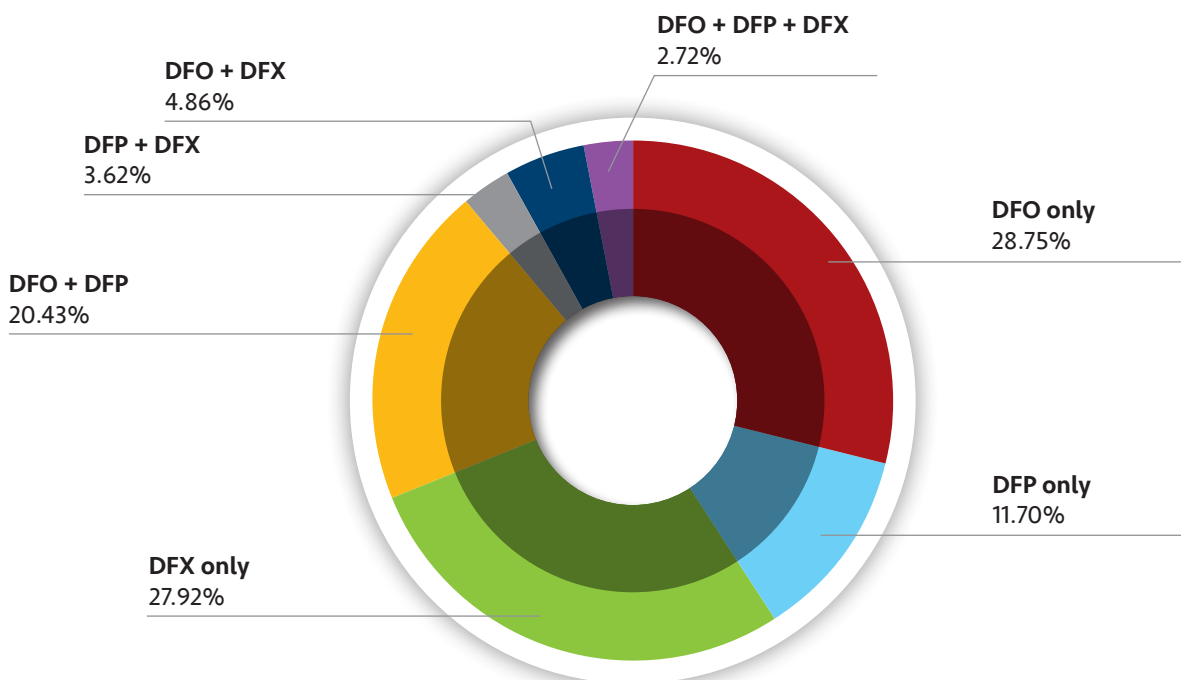
Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Tawau	34	DFO only	2	0.16
		DFP only	2	0.16
		DFX only	12	0.99
		DFO + DFP	8	0.66
		DFP + DFX	3	0.25
		DFO + DFX	4	0.33
		DFO + DFP + DFX	3	0.25
Keningau	129	DFO only	17	1.40
		DFP only	23	1.89
		DFX only	30	2.47
		DFO + DFP	29	2.39
		DFP + DFX	20	1.65
		DFO + DFX	6	0.49
		DFO + DFP + DFX	4	0.33
Tenom	20	DFO only	6	0.49
		DFP only	2	0.16
		DFX only	3	0.25
		DFO + DFP	7	0.58
		DFP + DFX	1	0.08
		DFO + DFX	1	0.08
		DFO + DFP + DFX	0	0.00
Beaufort	23	DFO only	7	0.58
		DFP only	3	0.25
		DFX only	10	0.82
		DFO + DFP	2	0.16
		DFP + DFX	0	0.00
		DFO + DFX	1	0.08
		DFO + DFP + DFX	0	0.00
Kudat	55	DFO only	9	0.74
		DFP only	2	0.16
		DFX only	21	1.73
		DFO + DFP	19	1.57
		DFP + DFX	0	0.00
		DFO + DFX	3	0.25
		DFO + DFP + DFX	1	0.08

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Kota Belud	22	DFO only	9	0.74
		DFP only	1	0.08
		DFX only	10	0.82
		DFO + DFP	1	0.08
		DFP + DFX	0	0.00
		DFO + DFX	1	0.08
		DFO + DFP + DFX	0	0.00
Ranau	60	DFO only	33	2.72
		DFP only	1	0.08
		DFX only	18	1.48
		DFO + DFP	6	0.49
		DFP + DFX	0	0.00
		DFO + DFX	1	0.08
		DFO + DFP + DFX	1	0.08
Lahad Datu	29	DFO only	7	0.58
		DFP only	3	0.25
		DFX only	10	0.82
		DFO + DFP	5	0.41
		DFP + DFX	1	0.08
		DFO + DFX	2	0.16
		DFO + DFP + DFX	1	0.08
Kota Marudu	98	DFO only	7	0.58
		DFP only	23	1.89
		DFX only	22	1.81
		DFO + DFP	41	3.38
		DFP + DFX	2	0.16
		DFO + DFX	3	0.25
		DFO + DFP + DFX	0	0.00
Sipitang	19	DFO only	2	0.16
		DFP only	2	0.16
		DFX only	9	0.74
		DFO + DFP	4	0.33
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	2	0.16

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Papar	17	DFO only	3	0.25
		DFP only	2	0.16
		DFX only	12	0.99
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Tambunan	32	DFO only	6	0.49
		DFP only	2	0.16
		DFX only	11	0.91
		DFO + DFP	6	0.49
		DFP + DFX	3	0.25
		DFO + DFX	2	0.16
		DFO + DFP + DFX	2	0.16
Beluran	16	DFO only	3	0.25
		DFP only	3	0.25
		DFX only	5	0.41
		DFO + DFP	5	0.41
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Semporna	2	DFO only	0	0.00
		DFP only	1	0.08
		DFX only	1	0.08
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Kinabatangan	25	DFO only	11	0.91
		DFP only	1	0.08
		DFX only	8	0.66
		DFO + DFP	2	0.16
		DFP + DFX	0	0.00
		DFO + DFX	2	0.16
		DFO + DFP + DFX	1	0.08

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
HWKKS	178	DFO only	28	2.31
		DFP only	24	1.98
		DFX only	92	7.58
		DFO + DFP	10	0.82
		DFP + DFX	6	0.49
		DFO + DFX	18	1.48
		DFO + DFP + DFX	0	0.00
Pitas	36	DFO only	19	1.57
		DFP only	2	0.16
		DFX only	10	0.82
		DFO + DFP	2	0.16
		DFP + DFX	3	0.25
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Kuala Penyu	11	DFO only	2	0.16
		DFP only	1	0.08
		DFX only	2	0.16
		DFO + DFP	2	0.16
		DFP + DFX	0	0.00
		DFO + DFX	4	0.33
		DFO + DFP + DFX	0	0.00
Kunak	3	DFO only	0	0.00
		DFP only	2	0.16
		DFX only	0	0.00
		DFO + DFP	1	0.08
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Tuaran	16	DFO only	2	0.16
		DFP only	0	0.00
		DFX only	12	0.99
		DFO + DFP	1	0.08
		DFP + DFX	1	0.08
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			1214	100.00

Figure 13.7: Distribution of Patients in Sabah by Type of Iron Chelator Received



222

Table 13.16: Distribution of Patients in Sabah According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0 - 4.9	63	DFO only	6	0.49
		DFP only	0	0.00
		DFX only	56	4.61
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.08
		DFO + DFP + DFX	0	0.00
5-9.9	245	DFO only	19	1.57
		DFP only	14	1.15
		DFX only	168	13.84
		DFO + DFP	7	0.58
		DFP + DFX	18	1.48
		DFO + DFX	17	1.40
		DFO + DFP + DFX	2	0.16
10-14.9	290	DFO only	52	4.28
		DFP only	55	4.53
		DFX only	101	8.32
		DFO + DFP	33	2.72
		DFP + DFX	20	1.65
		DFO + DFX	22	1.81
		DFO + DFP + DFX	7	0.58

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
15-19.9	238	DFO only	125	10.30
		DFP only	26	2.14
		DFX only	10	0.82
		DFO + DFP	62	5.11
		DFP + DFX	4	0.33
		DFO + DFX	9	0.74
		DFO + DFP + DFX	2	0.16
20-24.9	200	DFO only	91	7.50
		DFP only	10	0.82
		DFX only	4	0.33
		DFO + DFP	78	6.43
		DFP + DFX	0	0.00
		DFO + DFX	6	0.49
		DFO + DFP + DFX	11	0.91
25-29.9	96	DFO only	31	2.55
		DFP only	8	0.66
		DFX only	0	0.00
		DFO + DFP	45	3.71
		DFP + DFX	2	0.16
		DFO + DFX	2	0.16
		DFO + DFP + DFX	8	0.66
30-34.9	46	DFO only	15	1.24
		DFP only	13	1.07
		DFX only	0	0.00
		DFO + DFP	13	1.07
		DFP + DFX	0	0.00
		DFO + DFX	2	0.16
		DFO + DFP + DFX	3	0.25
35-39.9	21	DFO only	6	0.49
		DFP only	7	0.58
		DFX only	0	0.00
		DFO + DFP	8	0.66
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

13 Sabah

224

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
40=44.9	8	DFO only	2	0.16
		DFP only	4	0.33
		DFX only	0	0.00
		DFO + DFP	2	0.16
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-49.9	4	DFO only	1	0.08
		DFP only	3	0.25
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50-54.9	2	DFO only	1	0.08
		DFP only	1	0.08
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	1	DFO only	0	0.00
		DFP only	1	0.08
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60-64.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Above 65	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			1214	100.00

13.4.2 Serum Ferritin Level

From the limited data on ferritin level retrievable from MTR up to November 2018, number of patients in Sabah who had their serum ferritin level measured in 2018 was only 687 patients. A total of 133 patients (19.36%) have a serum ferritin level below 1000 ng/mL, 209 patients (30.42%) have a serum ferritin level between 1000-2499 ng/mL, and 167 patients (24.31%) have a serum ferritin level between 2500-4999 ng/mL. Patients with a serum ferritin level between 5000-9999 ng/mL numbers at 156 (22.71%), whereas number of patients with a serum ferritin level above 10000 ng/mL is 22 (3.20%).

Table 13.17: Distribution of Patients in Sabah According to Most Recent Serum Ferritin Level by Centre

Centre	Total	< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
Beaufort	20	5	0.73	5	0.73	6	0.87	4	0.58	0	0.00
Beluran	1	1	0.15	0	0.00	0	0.00	0	0.00	0	0.00
DOK	155	26	3.78	52	7.57	37	5.39	37	5.39	3	0.44
HWKKS	163	41	5.97	75	10.92	27	3.93	18	2.62	2	0.29
Keningau	10	1	0.15	1	0.15	6	0.87	2	0.29	0	0.00
Kinabatangan	26	0	0.00	3	0.44	11	1.60	12	1.75	0	0.00
Kota Belud	2	0	0.00	0	0.00	2	0.29	0	0.00	0	0.00
Kota Marudu	1	0	0.00	0	0.00	0	0.00	1	0.15	0	0.00
Kuala Penyu	14	5	0.73	3	0.44	4	0.58	2	0.29	0	0.00
Kudat	59	10	1.46	13	1.89	16	2.33	16	2.33	4	0.58
Kunak	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Lahad Datu	29	7	1.02	4	0.58	8	1.16	8	1.16	2	0.29
Papar	36	3	0.44	6	0.87	10	1.46	16	2.33	1	0.15
Pitas	22	3	0.44	3	0.44	9	1.31	7	1.02	0	0.00
HQE	32	7	1.02	8	1.16	6	0.87	8	1.16	3	0.44
Ranau	2	1	0.15	1	0.15	0	0.00	0	0.00	0	0.00
Semporna	2	0	0.00	2	0.29	0	0.00	0	0.00	0	0.00
Sipitang	16	2	0.29	4	0.58	5	0.73	3	0.44	2	0.29
Tambunan	29	7	1.02	6	0.87	8	1.16	6	0.87	2	0.29
Tawau	30	4	0.58	12	1.75	7	1.02	7	1.02	0	0.00

Serum Ferritin Level (ng/mL)	< 1000		1000-2499		2500-4999		5000-9999		10,000+		
	Centre	Total	No.	%	No.	%	No.	%	No.	%	No.
Tenom	20	3	0.44	5	0.73	3	0.44	6	0.87	3	0.44
Tuaran	18	7	1.02	6	0.87	2	0.29	3	0.44	0	0.00
Total	687	133	19.36	209	30.42	167	24.31	156	22.71	22	3.20

13.5 Observation and Comments

There are 22 new thalassaemia patients in Sabah in 2018, with 2 new thalassaemia births recorded. This is a reduction in number of new cases in Sabah compared to number of new cases in 2017. The highest prevalence of thalassaemia patients are seen at the northern regions of Sabah, such as in Kudat, Kota Marudu and Pitas (148.6 patients per 100,000), followed by the central region of Sabah (Kota Belud, Kota Kinabalu, Papar, Penampang, Putatan, Ranau and Tuaran) with 81.87 patients per 100,000. The majority of these patients are of Kadazan-Dusun ethnicity. Beta thalassaemia major constitutes the most common diagnosis in the state, leading Sabah to have the highest prevalence of the diagnosis in the whole country. The median patient age group is 10-14.9 years old, similar to 2017.

The serum ferritin levels of only 687 patients were successfully captured in the whole state of Sabah. This is a small improvement compared to 2017, where there were only 68 serum ferritin levels captured in the registry. However, there is still struggle with lack of appropriate funds to ensure a more consistent record of patients' serum ferritin levels in each hospital.

Only 49.78% of patients have a serum ferritin level below 2500 ng/mL. This is only a marginal improvement from 2017, which had 44.12% of patients with serum ferritin level below 2500 ng/mL. There is also struggle to improve serum ferritin levels in the patients, as 25.91% of patients have a serum ferritin higher than 5000 ng/mL.

There is a lot of incomplete data entered into the MTR. The main factor is that not all the patient case notes in each hospital have systematic and effective data management system to allow correct and relevant data to be easily retrieved. Data especially on blood transfusion, height, weight, and also serum ferritin level needs to be updated from time to time. Information regarding cause of death needs to be improved in the documentations, as a large group of patient are categorised under an "unknown" cause of death.

13.5.1 Recommendations

1. Definition and diagnosis of disease under the "others" category have to be standardised in all centres.
2. A more constant and stable funds for laboratory, especially for measuring serum ferritin levels, may help the measurements be done more regularly in the state.
3. The state struggles with managing thalassaemia patients in terms of adequately transfusing them and providing adequate iron chelation. Continuous patient education and early screening will help to overcome this problem.
4. Majority of patients in Sabah are poorly managed in hospitals with no specialists or paediatricians. Adequate resources are needed to conduct regular programmes to train the medical doctors and nurses in the districts. Furthermore, with a dedicated team, the management of thalassaemia patients in Sabah can be improved.

14 Sarawak

14.1 Introduction

In the 2018 census, the population of Sarawak is 2,829,300 people. Sarawak, also known as 'The Land of Hornbills', is situated northwest on the island of Borneo and is the largest state in Malaysia. Sarawak's population is very diverse, comprising many races and ethnic groups. There are 40 ethnic groups in Sarawak. Ibans form the largest group (29%), followed by Malays (23%), Chinese (22%), and other Bumiputera Sarawak (26%), namely Bidayuh (8%), Melanau (5%) and others (7%). Non-citizens make up 6% of population.

Sarawak has one tertiary hospital, which is Hospital Umum Sarawak and three regional hospitals, that is Hospital Sibu, Hospital Miri and Hospital Bintulu. Kapit, Sarikei and Sri Aman are district hospitals with specialists and the rest are district hospitals without specialists. Thalassaemia patients are seen at all these hospitals, with input from the specialists in general and regional hospitals.

As of November 2018, a total of 244 patients in Sarawak are registered in the MTR. Twelve of them have been cured by stem cell transplant and 21 patients have passed away since the registry was started in 1997.

Table 14.1: Population of Thalassaemia Patients in Sarawak by District

Administrative District	No. of Beta Thalassaemia Patients	No. of Alpha Thalassaemia Patients	Total Population
Kuching Division:	84	37	792,600
Kuching	81	36	693,800
Bau	1	0	60,700
Lundu	2	1	38,100
Samarahan Division:	2	7	199,300
Samarahan	2	6	99,800
Simunjan	0	1	45,500
Asajaya	0	0	54,000
Serian Division	6	0	103,100
Sri Aman Division:	0	3	108,100
Sri Aman	0	3	76,000
Lubok Antu	0	0	32,100
Betong Division:	3	1	124,100
Betong	2	0	71,300
Saratok	1	1	52,800
Sarikei Division	1	0	65,600
Sibu Division:	10	1	314,500
Kanowit	0	1	33,300
Sibu	10	0	281,200
Mukah Division	0	0	50,700
Bintulu Division:	6	5	259,100
Bintulu	6	5	223,200
Tatau	0	0	35,900

14 Sarawak

Administrative District	No. of Beta Thalassaemia Patients	No. of Alpha Thalassaemia Patients	Total Population
Kapit Division:	0	1	130,800
Belaga	0	0	43,200
Kapit	0	1	63,900
Song	0	0	23,700
Miri Division:	22	3	422,200
Miri	22	3	347,600
Marudi	0	0	74,600
Limbang Division:	28	3	142,900
Limbang	12	1	76,650
Lawas	16	2	66,250
Total	162	61	2,829,300

228

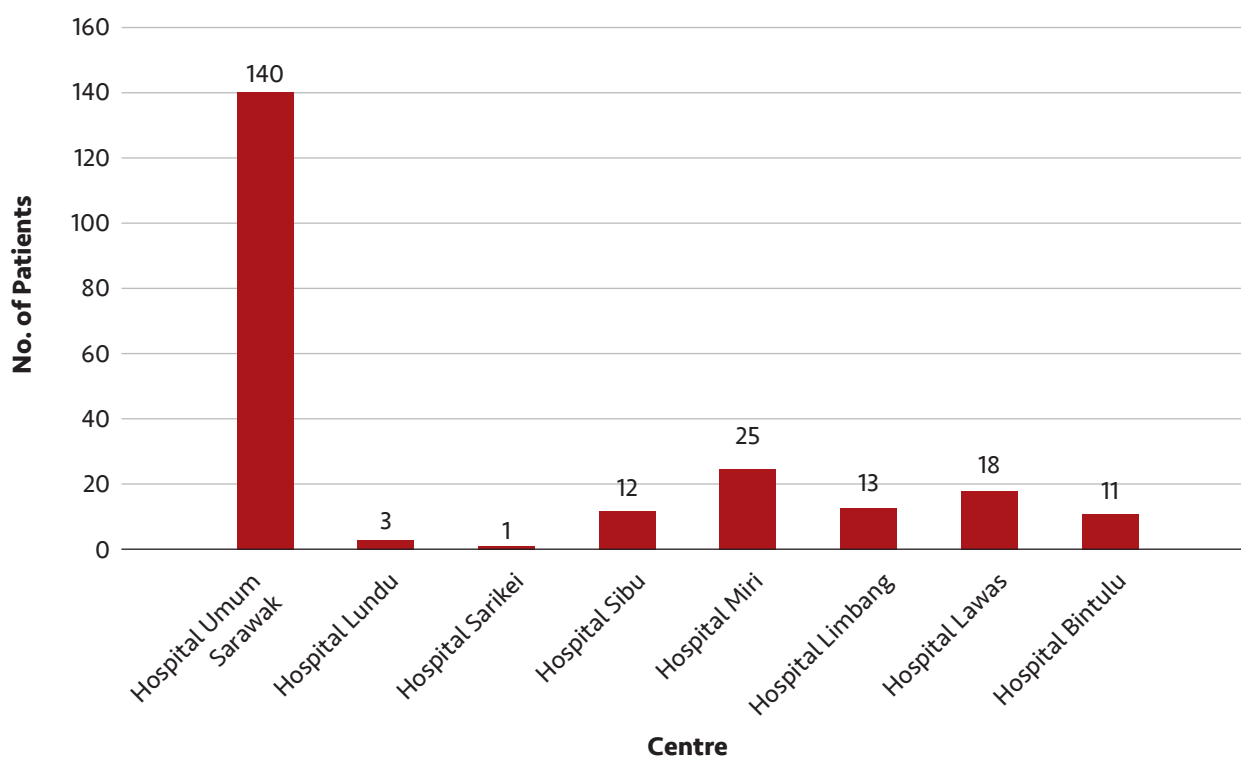
14.2 Patient Demographics

There are 223 living patients in Sarawak, including transplant patients. The main centre for treatment of thalassaemia patients in Sarawak is Hospital Umum Sarawak. This is followed by Hospital Miri, Hospital Lawas, Hospital Limbang, Hospital Sibul and Hospital Bintulu. Tables 14.2 and 14.3, and Figures 14.1 and 14.2 shows the distribution of thalassaemia patients in Sarawak.

Table 14.2: Distribution of Patients in Sarawak by Centre

Centre	No. of Patients (n)	Percentage (%)
Hospital Umum Sarawak	140	62.78
Hospital Lundu	3	1.35
Hospital Sarikei	1	0.45
Hospital Sibul	12 (1 Kapit)	5.38
Hospital Miri	25	11.21
Hospital Limbang	13	5.83
Hospital Lawas	18	8.07
Hospital Bintulu	11	4.93
Total	223	100.00

Figure 14.1: Distribution of Patients in Sarawak by Centre



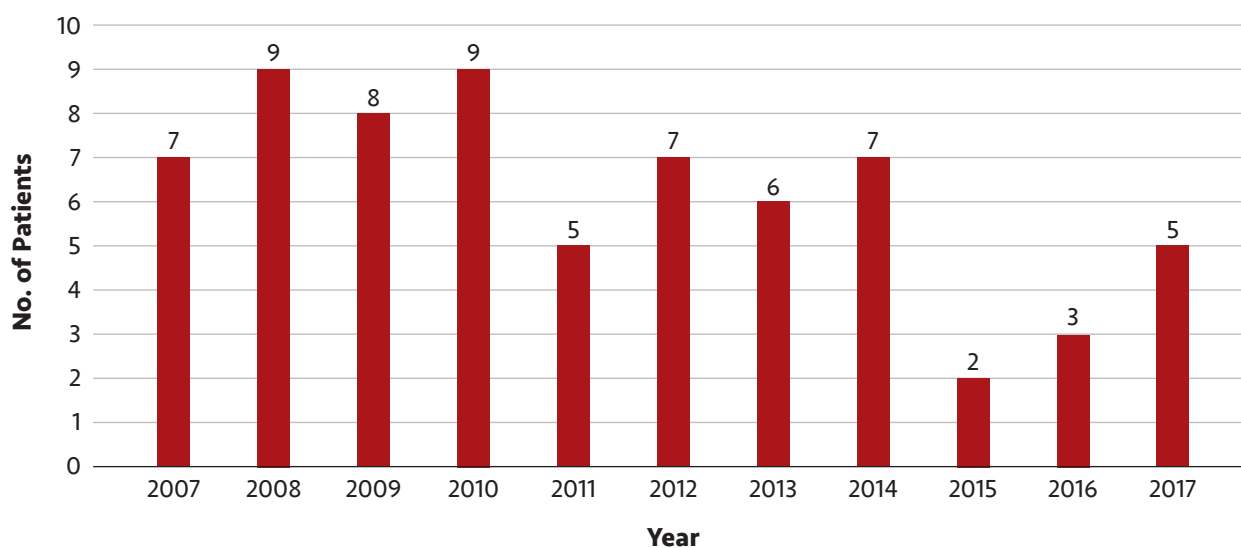
229

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Table 14.3: Distribution of Patients in Sarawak by Vital Status

Vital Status	No. Of Patients
Alive	206
Cumulative Reported Cured By Stem Cell Therapy	12
Lost To Follow-up	5
Total	223
Deaths in 2018	3
Cumulative Reported Deaths	21

Figure 14.2: New Thalassaemia Births in Sarawak by Year



14.2.1 Age

The youngest patients in Sarawak are 1 year old (two HbE-beta thalassaemia patients with the same date of birth) and eldest is 75 years old (HbH disease). The number of thalassaemia births in Sarawak has not shown significant decrease. The younger patients are mainly diagnosed with thalassaemia major. These patients' disease presentation was early, and they require regular transfusions and iron chelation therapy. Most of the older patients in Sarawak have HbH disease, and do not require any transfusions.

Table 14.4: Distribution of Patients in Sarawak by Age Group

Age Group (years)	No. of Beta Thalassaemia Patients	No. of Alpha Thalassaemia Patients	Total
0 - 4.9	13	4	17
5 - 9.9	22	12	34
10 -14.9	28	7	35
15 -19.9	26	7	33
20 - 24.9	34	8	42
25 - 29.9	13	3	16
30 - 34.9	12	9	21
35 - 39.9	5	4	9
40 - 44.9	2	0	2
45 - 49.9	2	4	6
50 - 54.9	1	3	4
55 - 59.9	0	1	1
60 - 64.9	1	0	1
Above 65	0	2	2
Total	159	64	223

Figure 14.3: Distribution of Patients in Sarawak by Age Group

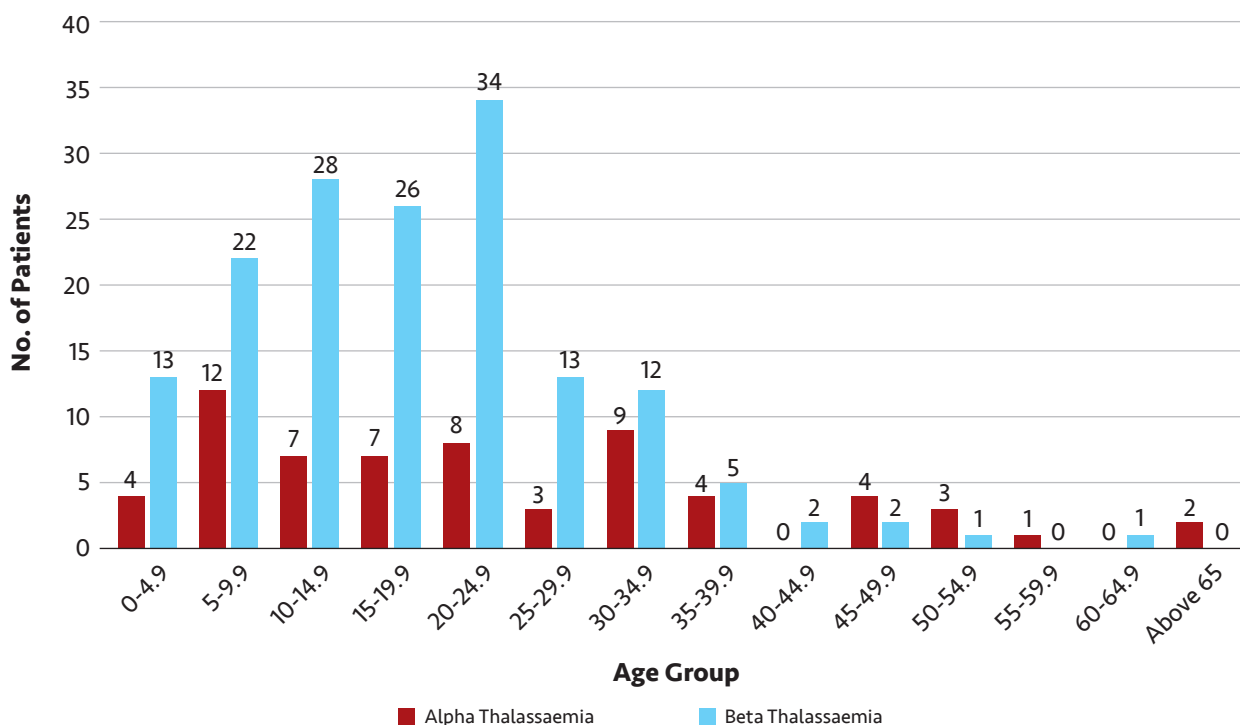
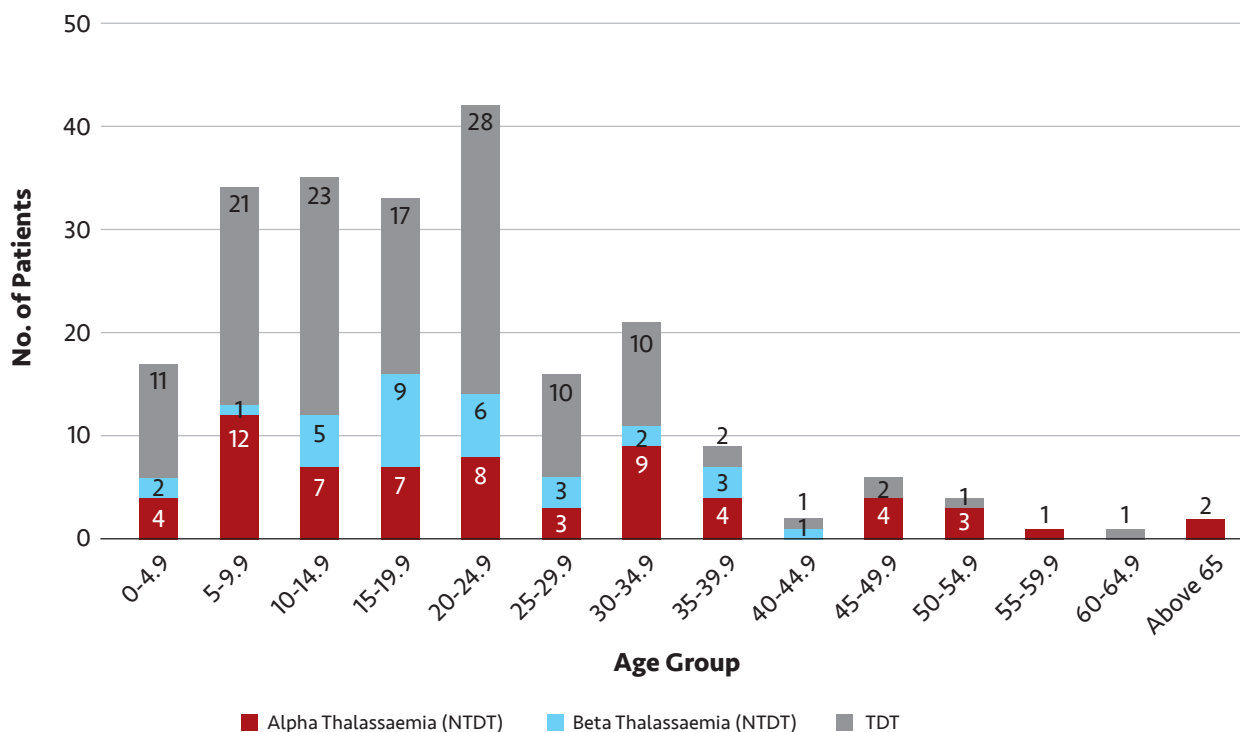


Table 14.5: Distribution of TDT and NTD (Alpha and Beta Thalassaemia) Patients in Sarawak by Age Group

Age Group (years)	No. of Patients (TDT)	No. of Patients (NTDT)		Total
		Alpha Thalassaemia	Beta Thalassaemia	
0 – 4.9	11	4	2	17
5 – 9.9	21	12	1	34
10 – 14.9	23	7	5	35
15 – 19.9	17	7	9	33
20 – 24.9	28	8	6	42
25 – 29.9	10	3	3	16
30 – 34.9	10	9	2	21
35 – 39.9	2	4	3	9
40 – 44.9	1	0	1	2
45 – 49.9	2	4	0	6
50 – 54.9	1	3	0	4
55 – 59.9	0	1	0	1
60 – 64.9	1	0	0	1
Above 65	0	2	0	2
Total	127	64	32	223

Figure 14.4: Distribution of TDT and NTD (Alpha and Beta Thalassaemia) Patients in Sarawak by Age Group



Five patients above the age of 40 were converted to TDT in their late 30s and they require transfusions every 2-3 months.

Median age of patients in Sarawak for both TDT and NTD is 15-19.9 years old.

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

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	17	Beta Thalassaemia Major	7	3.14
		Beta Thalassaemia Intermedia	1	0.45
		HbE-Beta Thalassaemia	5	2.24
		HbH Disease	4	1.79
		Others	0	0.00
5 - 9.9	34	Beta Thalassaemia Major	9	4.04
		Beta Thalassaemia Intermedia	3	1.35
		HbE-Beta Thalassaemia	10	4.48
		HbH Disease	10	4.48
		Others	2	0.90
10 -14.9	35	Beta Thalassaemia Major	15	6.73
		Beta Thalassaemia Intermedia	8	3.59
		HbE-Beta Thalassaemia	5	2.24
		HbH Disease	6	2.69
		Others	1	0.45
15 -19.9	33	Beta Thalassaemia Major	17	7.62
		Beta Thalassaemia Intermedia	2	0.90
		HbE-Beta Thalassaemia	7	3.14
		HbH Disease	6	2.69
		Others	1	0.45
20 - 24.9	42	Beta Thalassaemia Major	23	10.31
		Beta Thalassaemia Intermedia	4	1.79
		HbE-Beta Thalassaemia	7	3.14
		HbH Disease	8	3.59
		Others	0	0.00
25 - 29.9	16	Beta Thalassaemia Major	10	4.48
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	3	1.35
		HbH Disease	1	0.45
		Others	2	0.90
30 - 34.9	21	Beta Thalassaemia Major	11	4.93
		Beta Thalassaemia Intermedia	1	0.45
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	8	3.59
		Others	1	0.45

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	9	Beta Thalassaemia Major	2	0.90
		Beta Thalassaemia Intermedia	1	0.45
		HbE-Beta Thalassaemia	2	0.90
		HbH Disease	3	1.35
		Others	1	0.45
40 - 44.9	2	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	2	0.90
		HbH Disease	0	0.00
		Others	0	0.00
45 - 49.9	6	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.45
		HbE-Beta Thalassaemia	1	0.45
		HbH Disease	4	1.79
		Others	0	0.00
50 - 54.9	4	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.45
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	3	1.35
		Others	0	0.00
55 - 59.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.45
		Others	0	0.00
60 - 64.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.45
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	2	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	2	0.90
		Others	0	0.00
Total			223	100.00

233

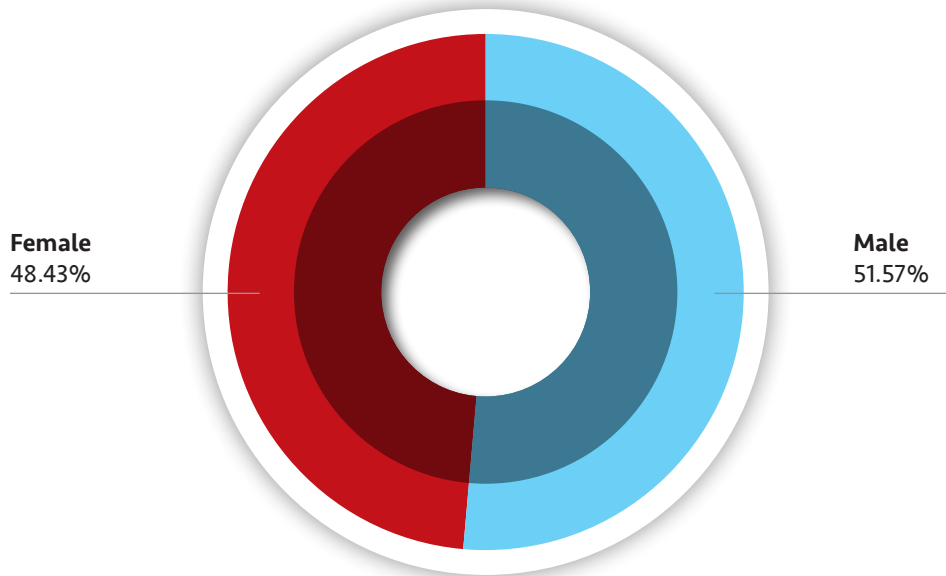
MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

14 Sarawak

14.2.2 Gender

The gender distribution of thalassaemia patients in Sarawak are as follows: 115 (51.57%) of patients are male and 108 (48.43%) of the patients are female.

Figure 14.5: Distribution of Patients in Sarawak by Gender



234

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

Centre	Male		Female	
	No.	%	No.	%
Hospital Umum Sarawak	65	29.15	76	34.08
Hospital Lundu	2	0.90	1	0.45
Hospital Sarikei	0	0.00	1	0.45
Hospital Sibul	5	2.24	6	2.69
Hospital Miri	16	7.17	9	4.04
Hospital Limbang	10	4.48	3	1.35
Hospital Lawas	11	4.93	7	3.14
Hospital Bintulu	6	2.69	5	2.24
Total	115	51.57	108	48.43

14.3 Diagnosis

42.15% of the patients in Sarawak were diagnosed with beta thalassaemia major. This is followed by HbH disease (25.11%), HbE-beta thalassaemia (18.83%) and thalassaemia intermedia (10.31%). HbH disease is not properly reflected in this data as there is no screening done and patients are incidentally noted to be pale. Most of the HbH patients are asymptomatic.

Table 14.8: Distribution of Patients in Sarawak by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	94	42.15
Beta Thalassaemia Intermedia	23	10.31
HbE-Beta Thalassaemia	42	18.83
HbH Disease	56	25.11
Others	8	3.59
Total	223	100.00

Figure 14.6: Distribution of Patients in Sarawak by Diagnosis

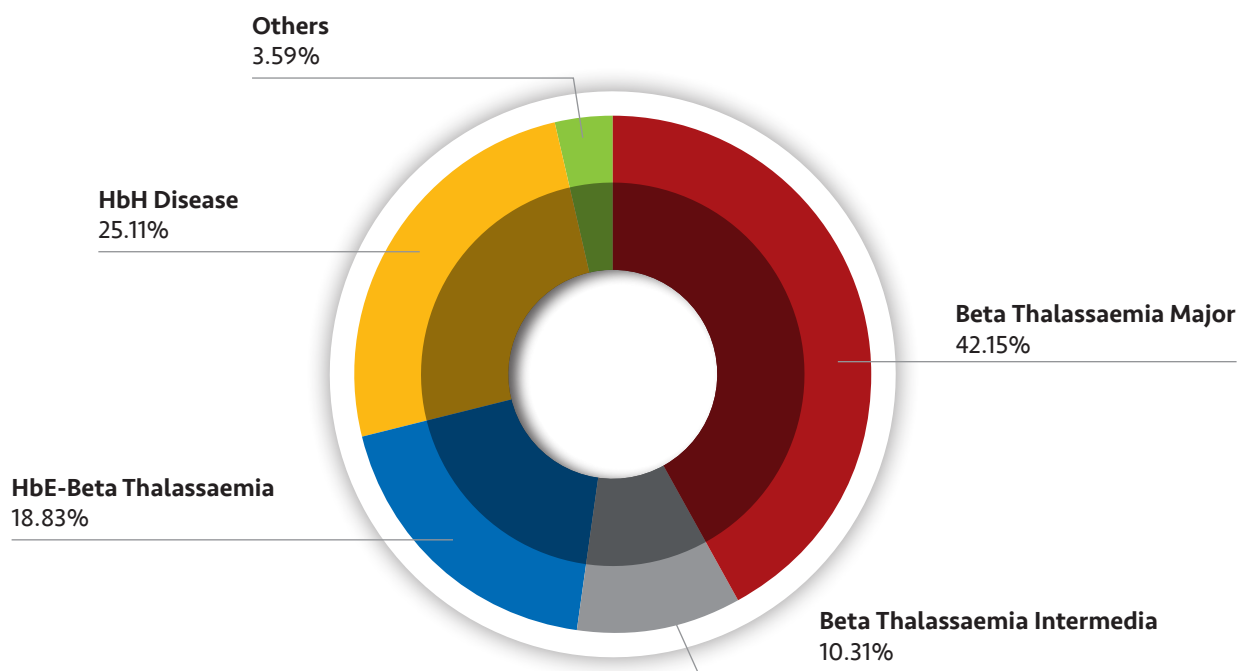


Table 14.9: Distribution of Patients in Sarawak According to Diagnosis by Centre

Diagnosis	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others		
	Centre	No.	%	No.	%	No.	%	No.	%	No.	%
HUS Kuching		46	20.63	11	4.93	35	15.7	44	19.73	5	2.24
Hospital Lundu		2	0.90	0	0.00	0	0.00	0	0.00	1	0.45
Hospital Sarikei		0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Sibul		10	4.48	1	0.45	0	0.00	1	0.45	0	0.00
Hospital Miri		16	7.17	5	2.24	0	0.00	3	1.35	1	0.45
Hospital Limbang		11	4.93	0	0.00	0	0.00	2	0.90	0	0.00
Hospital Lawas		6	2.69	4	1.79	6	2.69	2	0.90	0	0.00
Hospital Bintulu		3	1.35	2	0.90	1	0.45	4	1.79	1	0.45
Total		94	42.15	23	10.31	42	18.83	56	25.11	8	3.59

236

Table 14.10: Distribution of Patients in Sarawak According to Diagnosis by Ethnic Group

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	94	Malay	27	12.11
		Chinese	51	22.87
		Iban	0	0.00
		Bidayuh	1	0.45
		Kedayan	6	2.69
		Pribumi Sarawak	5	2.24
		Kadazan-Dusun	4	1.79
Beta Thalassaemia Intermedia	23	Malay	9	4.04
		Chinese	8	3.59
		Iban	0	0.00
		Bidayuh	0	0.00
		Kedayan	4	1.79
		Pribumi Sarawak	1	0.45
		Kadazan-Dusun	1	0.45
HbE-Beta Thalassaemia	42	Malay	35	15.70
		Chinese	5	2.24
		Iban	1	0.45
		Bidayuh	1	0.45
HbH Disease	56	Malay	19	8.52
		Chinese	21	9.42
		Iban	11	4.93
		Bidayuh	2	0.90
		Melanau	1	0.45
		Pribumi Sarawak	2	0.90

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Others	8	Malay	5	2.24
		Chinese	1	0.45
		Iban	2	0.90
Total			223	100.00

14.4 Treatment

14.4.1 Iron Chelation Therapy

The number of patients in Sarawak who are on iron chelation therapy is 124. A large group of patients are on DFX (45.16%). All young, newly diagnosed patients were started on DFX monotherapy by two years of age if ferritin level is higher than 1000 ng/mL. They were only switched to other drugs if they had side effects or could not tolerate the drug. Approximately 21.77% were on a combination of DFO and DFP, and 15.32% of patients are on DFP monotherapy (usually older patients or those who were initially on combination therapy, and when the serum ferritin levels dropped, continued on with DFP monotherapy). In addition, 14.52% of patients are on DFO monotherapy and 3.23% are receiving a combination of DFO and DFX.

Patients on combination therapy had poor MRI T2* or high serum ferritin level, despite optimal treatment with subcutaneous DFO.

Seven NTD patients in Sarawak required chelation. Three of these seven patients had fewer than five transfusions, and the rest were never transfused before. One patient started chelation therapy at age 12, while the others began after the age of 20 when their ferritin levels reached above 800 ng/mL.

Table 14.11: Distribution of Patients in Sarawak by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	18	14.52
DFP only	19	15.32
DFX only	56	45.16
DFO + DFP	27	21.77
DFP + DFX	0	0.00
DFO + DFX	4	3.23
DFO + DFP + DFX	0	0.00
Total	124	100.00

Table 14.12: Distribution of Patients in Sarawak According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
HUS, Kuching	68	DFO only	6	4.84
		DFP only	14	11.29
		DFX only	27	21.77
		DFO + DFP	17	13.71
		DFP + DFX	0	0.00
		DFO + DFX	4	3.23
		DFO + DFP + DFX	0	0.00
Hospital Lundu	2	DFO only	2	1.61
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Sibü	10	DFO only	4	3.23
		DFP only	0	0.00
		DFX only	5	4.03
		DFO + DFP	1	0.81
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Miri	17	DFO only	3	2.42
		DFP only	2	1.61
		DFX only	10	8.06
		DFO + DFP	2	1.61
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Limbang	9	DFO only	1	0.81
		DFP only	2	1.61
		DFX only	6	4.84
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Lawas	12	DFO only	2	1.61
		DFP only	1	0.81
		DFX only	3	2.42
		DFO + DFP	6	4.84
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Bintulu	6	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	5	4.03
		DFO + DFP	1	0.81
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			124	100.00

Figure 14.7: Distribution of Patients in Sarawak by Type of Iron Chelator Received

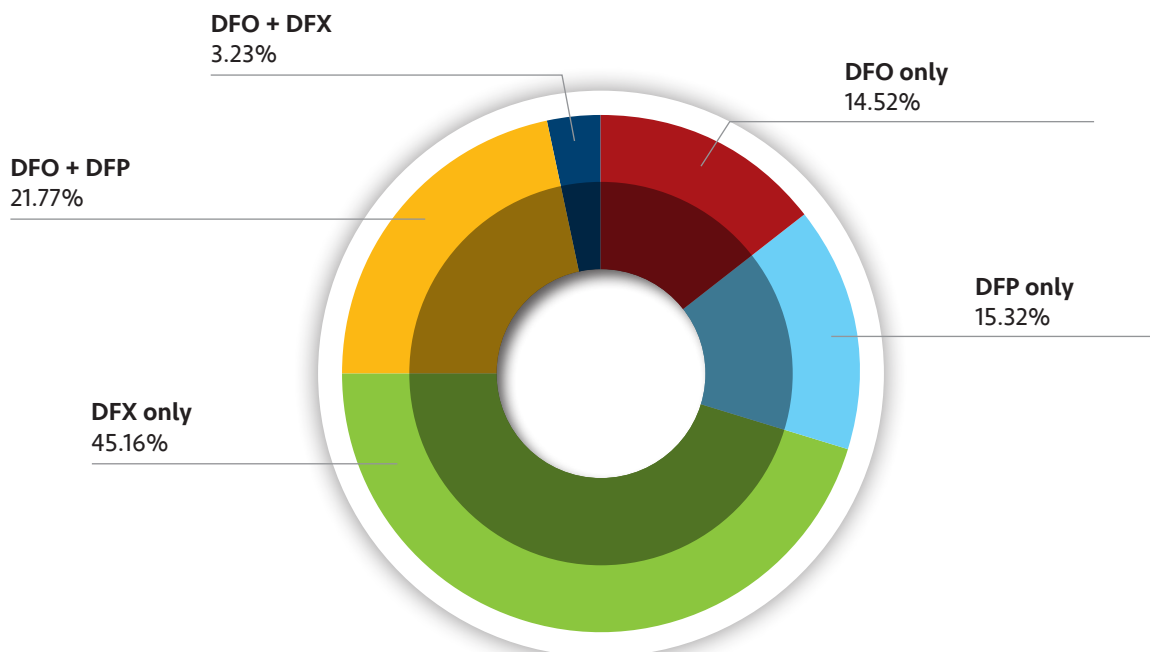


Table 14.13: Distribution of Patients in Sarawak According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	4	DFO only	1	0.81
		DFP only	0	0.00
		DFX only	3	2.42
		DFO + DFP	0	0.00
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
5-9.9	16	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	16	12.90
		DFO + DFP	0	0.00
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
10-14.9	24	DFO only	1	0.81
		DFP only	0	0.00
		DFX only	22	17.74
		DFO + DFP	1	0.81
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
15-19.9	22	DFO only	6	4.84
		DFP only	5	4.03
		DFX only	3	2.42
		DFO + DFP	8	6.45
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
20-24.9	25	DFO only	5	4.03
		DFP only	4	3.23
		DFX only	2	1.61
		DFO + DFP	12	9.68
		DFO + DFX	0	0.00
		DFP + DFX	2	1.61
25-29.9	12	DFO only	3	2.42
		DFP only	0	0.00
		DFX only	3	2.42
		DFO + DFP	5	4.03
		DFO + DFX	0	0.00
		DFP + DFX	1	0.81

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
30-34.9	11	DFO only	2	1.61
		DFP only	3	2.42
		DFX only	5	4.03
		DFO + DFP	1	0.81
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
35-39.9	4	DFO only	0	0.00
		DFP only	3	2.42
		DFX only	1	0.81
		DFO + DFP	0	0.00
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
40-44.9	1	DFO only	0	0.00
		DFP only	1	0.81
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
45-49.9	3	DFO only	0	0.00
		DFP only	1	0.81
		DFX only	1	0.81
		DFO + DFP	0	0.00
		DFO + DFX	1	0.81
		DFP + DFX	0	0.00
50-54.9	1	DFO only	0	0.00
		DFP only	1	0.81
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
55-59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
60-64.9	1	DFO only	0	0.00
		DFP only	1	0.81
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
Above 65	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFO + DFX	0	0.00
		DFP + DFX	0	0.00
Total			124	100.00

14.4.2 Serum Ferritin Level

A total of 146 patients in Sarawak have their serum ferritin level recorded in 2018. Of these, 100 (68.49%) are TDT patients and 46 (31.51%) are NTD patients. Sixty-three (43.15%) patients have a serum ferritin lower than 1000 ng/mL, 47 patients (32.19%) with serum ferritin level between 1000-2499 ng/mL, and 15 patients (10.27%) have very high serum ferritin level of above 5000 ng/mL.

Majority of patients (99 out of 110 patients, 90.0%) from regional centres and tertiary centres have a ferritin level lower than 2500 ng/mL, while 3 out of 36 patients (36.11%) from district hospitals have a ferritin level above 2500 ng/mL.

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

Centre	Total	< 1000		1000-2499		2500-4999		5000-9999		10,000+	
		No.	%	No.	%	No.	%	No.	%	No.	%
HUS, Kuching	88	47	32.19	22	15.07	14	9.59	4	2.74	1	0.68
Hospital Miri	19	9	6.16	8	5.48	2	1.37	0	0.00	0	0.00
Hospital Sibul	8	2	1.37	5	3.42	0	0.00	1	0.68	0	0.00
Hospital Bintulu	7	1	0.68	5	3.42	0	0.00	0	0.00	1	0.68
Hospital Lawas	12	3	2.05	3	2.05	1	0.68	4	2.74	1	0.68
Hospital Limbang	11	1	0.68	4	2.74	4	2.74	1	0.68	1	0.68
Hospital Lundu	1	0	0.00	0	0.00	0	0.00	1	0.68	0	0.68
Total	146	63	43.15	47	32.19	21	14.38	11	7.53	4	2.74

Figure 14.8: Distribution of Patients in Sarawak According to Most Recent Serum Ferritin Level

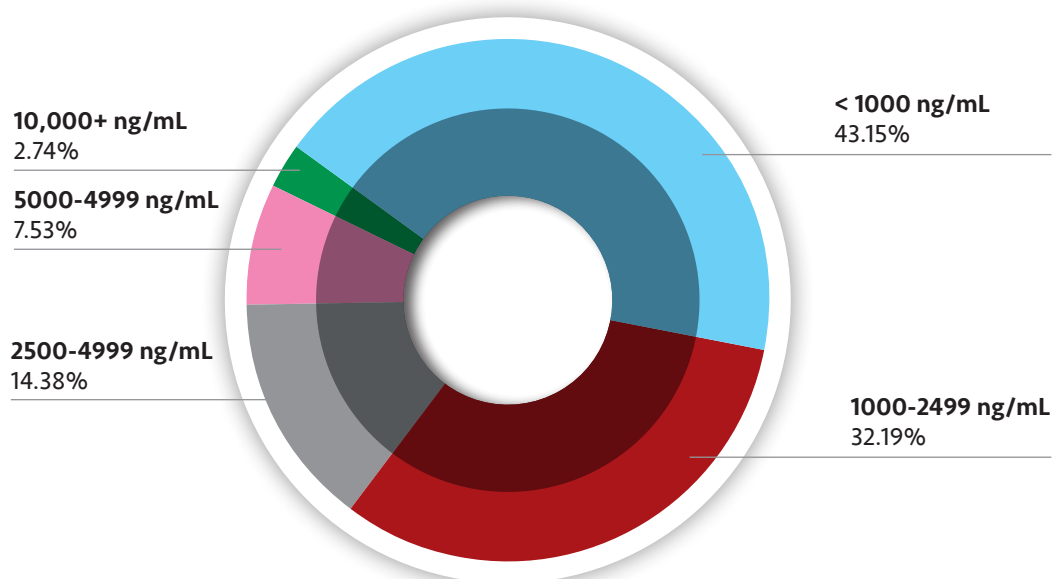


Table 14.15: Distribution of TDT Patients in Sarawak According to Most Recent Serum Ferritin Level by Age Group

Serum Ferritin Level (ng/mL)	Age Group (years)	Total	< 1000		1000-2499		2500-4999		5000-9999		10,000+	
			No.	%	No.	%	No.	%	No.	%	No.	%
0 - 4.9	7	2	2.00	3	3.00	2	2.00	0	0.00	0	0.00	
5 - 9.9	15	5	5.00	8	8.00	2	2.00	0	0.00	0	0.00	
10 - 14.9	21	5	5.00	14	14.00	1	1.00	1	1.00	0	0.00	
15 - 19.9	16	3	3.00	8	8.00	3	3.00	2	2.00	0	0.00	
20 - 24.9	20	5	5.00	4	4.00	6	6.00	4	4.00	1	1.00	
25 - 29.9	10	3	3.00	1	1.00	3	3.00	2	2.00	1	1.00	
30 - 34.9	5	2	2.00	1	1.00	1	1.00	0	0.00	1	1.00	
35 - 39.9	3	3	3.00	0	0.00	0	0.00	0	0.00	0	0.00	
Above 40	3	1	1.00	0	0.00	1	1.00	1	1.00	0	0.00	
Total	100	29	29.00	39	39.00	19	19.00	10	10.00	3	3.00	

Twenty-two patients with TDT in Sarawak are below 10 year old.

Sixty-eight patients (68%) have a ferritin level lower than 2500 ng/mL, of which 29 patients (29%) have a ferritin level lower than 1000 ng/mL. No patient below 10 years old have a ferritin level above 5000 ng/mL. This might mean a better outcome in later years if these patients continue to remain compliant.

Table 14.16: Distribution of NTD T Patients in Sarawak According to Most Recent Serum Ferritin Level by Age Group

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Age Group (years)	Total	No.	%	No.	%	No.	%	No.	%	No.	%
0 - 4.9	4	4	8.70	0	0.00	0	0.00	0	0.00	0	0.00
5 - 9.9	10	9	19.57	1	2.17	0	0.00	0	0.00	0	0.00
10 - 14.9	6	5	10.87	1	2.17	0	0.00	0	0.00	0	0.00
15 - 19.9	6	6	13.04	0	0.00	0	0.00	0	0.00	0	0.00
20 - 24.9	4	2	4.35	0	0.00	1	2.17	1	2.17	0	0.00
25 - 29.9	2	1	2.17	1	2.17	0	0.00	0	0.00	0	0.00
30 - 34.9	6	2	4.35	4	8.70	0	0.00	0	0.00	0	0.00
35 - 39.9	3	1	2.17	1	2.17	1	2.17	0	0.00	1	2.17
Above 40	5	4	8.70	1	2.17	0	0.00	0	0.00	0	0.00
Total	46	34	73.91	9	17.39	2	4.35	1	2.17	1	2.17

Majority of NTD T patients have a ferritin level below 1000 ng/mL. Only six patients with NTD T are on chelation therapy—four patients on DFP and two patients on DFX.

Table 14.17: Distribution of Patients in Sarawak According to Transfusion Status by Centre

Centre	No. of Patients (TDT)	No. of Patients (NTDT)
Hospital Umum Sarawak, Kuching	58	82
Hospital Lundu	2	1
Hospital Sarikei	1	0
Hospital Sibul	9	3
Hospital Miri	16	9
Hospital Limbang	12	1
Hospital Lawas	11	7
Hospital Bintulu	8	3
Total	117	106

14.5 Complications and Deaths

There were no new infectious complications in 2018 in Sarawak. This might mean safer blood are being given to patients.

Table 14.18: Infectious Complications in Thalassaemia Patients Above 15 Years Old in Sarawak

Infections	No. of Patients
Hepatitis B	1
Hepatitis C	7
HIV	0

There are 65 TDT patients who are above 15 years old in Sarawak.

31 out of 65 patients (47.69%) have delayed puberty and need some form of hormonal replacement. In addition, 30 patients (46.15%) have short stature.

Of the 19 patients who underwent the dual-energy X-ray absorptiometry (DEXA) scan, 84.21% showed evidence of either osteopenia or osteoporosis. This might have been contributed by the fact that their delayed puberty were not treated in a timely manner.

Sixty-nine patients have had their T2* done between 2015-2018. Of these, 82.6% have a normal T2*. Some of these patients had shown an initial T2* of lesser than 20 ms but had improved over the years with intensive chelation. Only 25 T2* scans were performed in 2018 (T2* scans were performed by a paediatric cardiologist, who does an echocardiography and check for pulmonary hypertension at the same time).

Out of the 25 scans performed in 2018, five poorly chelated and non-compliant patients had a T2* of lower than 10 ms. Four patients had a T2* of between 10-20 ms. These patients had delayed puberty, whereas the rest were normal.

Table 14.19: Endocrine Complications in Patients Above 15 Years Old in Sarawak Receiving Iron Chelation Therapy

Endocrine	No. of Patients
Delayed Puberty	31 (3 post BMT)
DM	5 (2 died, 1 post BMT)
Hypothyroidism	2
Growth Retardation	30
Bone Health (DEXA Scan)	No. of Patients
Osteoporosis	16
Normal DEXA scan	3

Table 14.20: Cardiac Complication in Patients in Sarawak

Cardiac (MRI T2*)	No. of Patients
< 10ms	5
10-20 ms	7
> 20ms	57

Sixty-nine patients had MRI T2 done between 2015-2018.*

Table 14.21: Causes of Death of Patients in Sarawak

Causes of Death	No. of Patients
Infections	4
Cardiac	14
Anaemia	1 (auto and allommunisation)
Old Age	1 (died at age of 88 years old)
Thrombosis	1
Total	21

There are 21 thalassaemia patient deaths in Sarawak since 1997. Three of these patients died in 2018 (two of infections, one due to cardiac complication).

14.6 Observation and Comments

From the data collection, there was an 8% increment in the number of thalassaemia patients in Sarawak. Two were TDT patients, and the rest were detected in clinics or family screening and only registered in 2018.

Approximately 75.34% of patients have a serum ferritin level lower than 2500 ng/mL, which is an improvement from only 15.77% in 2017, although this can be further improved on. Treatment centres without paediatricians face the biggest problem with iron chelator compliance. Both these centres are located at the northern region of Sarawak in Limbang and Lawas. This problem may be caused by no proper continuation of care due to high staff turnover. This problem remains regardless of whether method of chelation is by subcutaneous infusion or oral.

Treatment regime in Sarawak follows the clinical practice guideline (CPG) treatment protocol, i.e. using DFO as first line. Combination therapy is used when serum ferritin level does not drop even when at optimal DFO intake or if there are cardiac complications. DFX is limited to thalassaemia patients below 12 years old and to adults who are extremely non-compliant to DFO, due of financial constraints.

Data showed that the median age of thalassaemia patients in Sarawak is between 15-19.9 years old, which is the same as 2017. There are also more adult patients diagnosed in their 20s, mainly with alpha and HbE-beta thalassaemia.

As a significant number of patients were not well-chelated when they were younger, have a substantial number of endocrine complications, especially affecting growth and development. Sixty-five of TDT patients are above 15 years old. Out of these, approximately 50% have delayed puberty and short stature. Bone health also remains a major problem. There is also a need for safer blood with better infectious screening, as 3.14% of patients have hepatitis C. MRI T2* scans were attempted on all chelated patients above 10 years of age. Out of the 69 patients who had a T2* scan done, 17.39% have MRI T2* below 20 ms.

Since the beginning of the registry, Sarawak has recorded 21 deaths, mostly due to cardiac complications. Three deaths were recorded in 2018; one death was due to cardiac complication (severely non-compliant patient) and two from infections.

14.6.1 Recommendations:

1. There is a need to liaise with the haemato-pathologists/polyclinics to inform the clinical side once they have a diagnosis of thalassaemia intermedia/HbH disease. This hopefully will have a better pickup rate and give an actual number.
2. To improve treatment outcomes in Northern Sarawak. There is plan to intensify training in these sites and to assign a paediatrician to Limbang, to improve compliance and outcome in these sites.
3. Safer blood is required in Sarawak. NAT testing should be brought into Sarawak to reduce risk of infections.
4. To reduce risk of osteoporosis, delayed puberty should be treated in a timely manner.
5. Healthcare providers must keep the infectious complications in mind, and to stop chelation therapy if patients are unwell. Admit for antibiotics and watch for neutropenia, especially if patients are on DFP or are splenectomised.

15 Selangor

15.1 Introduction

Selangor is one of the 13 states in Malaysia. It is the eighth largest state in Peninsular Malaysia with an estimated population of 6,470,000 in 2018. Selangor is divided into 9 administrative districts, which are Gombak, Hulu Langat, Hulu Selangor, Klang, Kuala Langat, Kuala Selangor, Petaling, Sabak Bernam and Sepang.

There are 1022 living thalassaemia patients in Selangor. Most thalassaemia patients in the state receive transfusions and follow-up care in the hospitals listed in Table 15.1. Most hospitals carry out transfusions in their ACC. Hospitals without an ACC carry out transfusions in the wards.

15.2 Patient Demographics

A total of 1169 thalassaemia patients were recorded in Selangor. These patients receive transfusion and care at the 11 centres in Table 15.1. Hospital Ampang has the majority of patients in Selangor, as all paediatric patients from the Institute of Paediatrics, Hospital Kuala Lumpur (IPKHL) and Hospital Selayang are transferred to this centre when they turn 18 years old. Hospital Tengku Ampuan Rahimah (HTAR), Klang is the centre of referral for Selangor Central Zone hospital cluster and has the second highest number of patients in Selangor.

Table 15.1: Distribution of Patients in Selangor by Centre

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
Hospital Ampang	703	60.14
Hospital Banting	16	1.37
Hospital Kajang	43	3.68
Hospital Kuala Kubu Bharu	3	0.26
Hospital Selayang	82	7.01
Hospital Serdang	35	2.99
Hospital Shah Alam	13	1.11
Hospital Sungai Buloh	63	5.39
Hospital Tanjung Karang	3	0.26
Hospital Tengku Ampuan Jemaah, Sabak Bernam	7	0.60
Hospital Tengku Ampuan Rahimah, Klang	201	17.19
Total	1169	100.00

15 Selangor

Figure 15.1: Distribution of Patients in Selangor by Centre

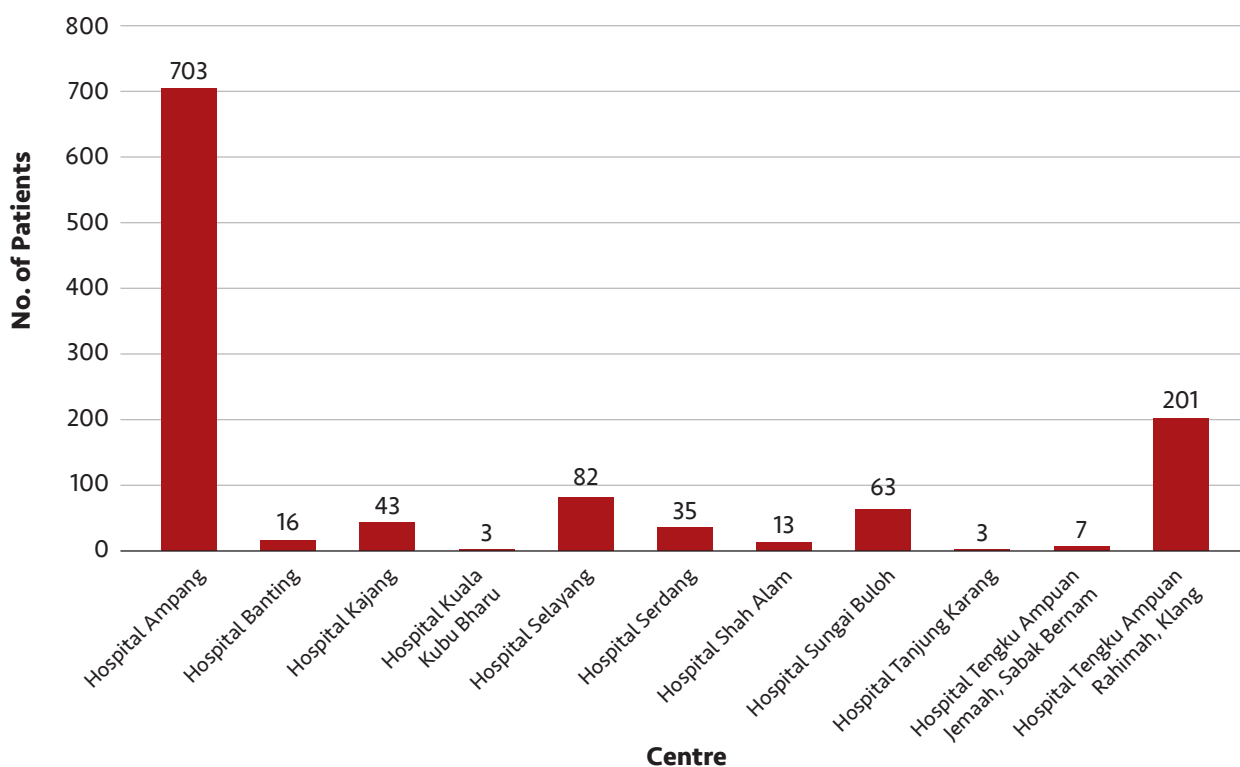


Table 15.2: Distribution of Patients in Selangor by Vital Status

Vital Status	Patients
Alive	1022
Cumulative Reported Cured by Stem Cell Therapy	10
Lost to Follow-up	137
Total	1169
Deaths in 2018	10
Cumulative Reported Deaths	64

Table 15.3: Cumulative Causes of Death Since 1997 in Selangor

Cause of Death	No. of Patients
Infections	20
Cardiac Causes	22
Liver Disease	4
Tumours	2
Others	13
Unknown*	3
Total	64

*Data not available, patient died at home.

15.2.1 Age

Approximately 35.10% of thalassaemia patients in Selangor are below 20 years old, falling under paediatric and adolescent age categories. Adult patients above 21 years old forms 64.93% of the thalassaemia cases. The oldest patient in Selangor is an 86-year-old diagnosed with beta thalassaemia intermedia.

Table 15.4: Distribution of Patients in Selangor by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 – 4.9	54	4.62
5 – 9.9	119	10.18
10 – 14.9	124	10.61
15 – 19.9	113	9.67
20 – 24.9	165	14.11
25 – 29.9	139	11.89
30 – 34.9	142	12.15
35 – 39.9	104	8.90
40 – 44.9	84	7.19
45 – 49.9	42	3.59
50 – 54.9	25	2.14
55 – 59.9	17	1.45
60 – 64.9	17	1.45
Above 65	24	2.05
Total	1169	100.00

249

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Figure 15.2: Distribution of Patients in Selangor by Age Group

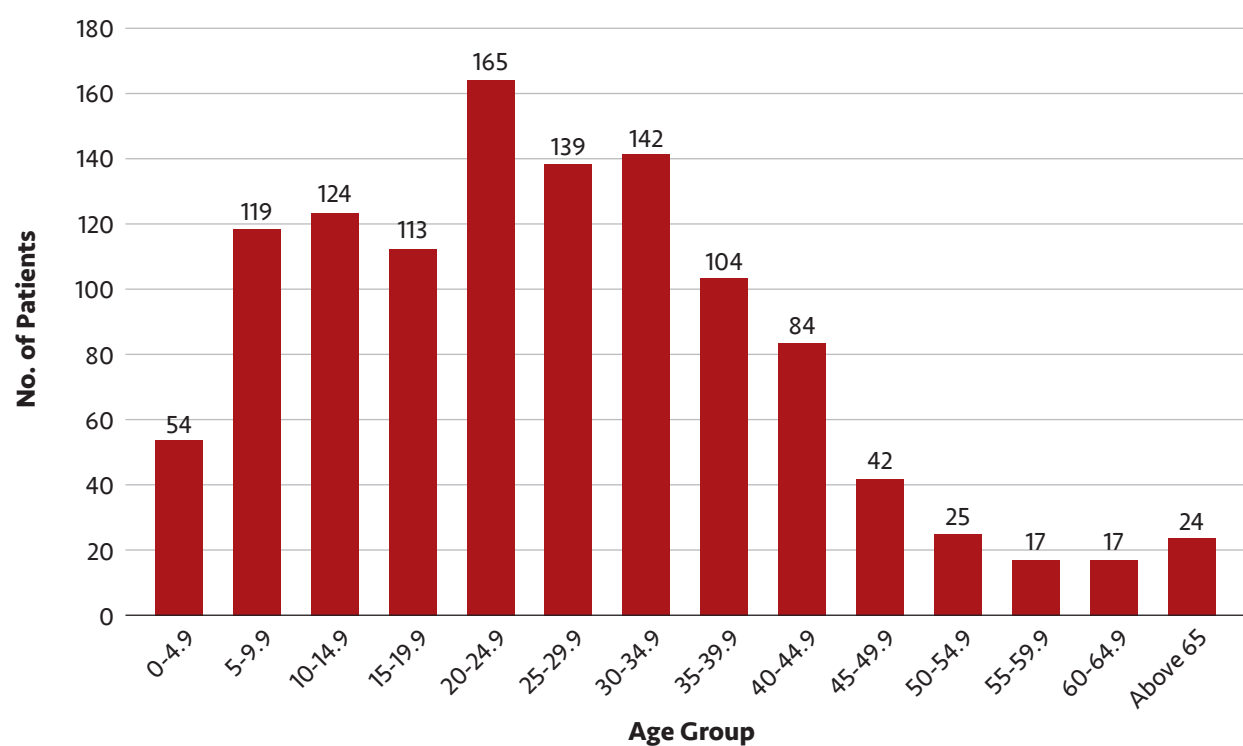


Table 15.5: Distribution of Patients in Selangor According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	54	Beta Thalassaemia Major	15	1.28
		Beta Thalassaemia Intermedia	25	2.14
		HbE-Beta Thalassaemia	2	0.17
		HbH Disease	9	0.77
		Others	3	0.26
5 - 9.9	119	Beta Thalassaemia Major	23	1.97
		Beta Thalassaemia Intermedia	56	4.79
		HbE-Beta Thalassaemia	8	0.68
		HbH Disease	19	1.63
		Others	13	1.11
10 -14.9	124	Beta Thalassaemia Major	19	1.63
		Beta Thalassaemia Intermedia	70	5.99
		HbE-Beta Thalassaemia	10	0.86
		HbH Disease	18	1.54
		Others	7	0.60
15 -19.9	113	Beta Thalassaemia Major	19	1.63
		Beta Thalassaemia Intermedia	63	5.39
		HbE-Beta Thalassaemia	8	0.68
		HbH Disease	21	1.80
		Others	2	0.17
20 - 24.9	165	Beta Thalassaemia Major	55	4.70
		Beta Thalassaemia Intermedia	60	5.13
		HbE-Beta Thalassaemia	8	0.68
		HbH Disease	37	3.17
		Others	5	0.43
25 - 29.9	139	Beta Thalassaemia Major	28	2.40
		Beta Thalassaemia Intermedia	66	5.65
		HbE-Beta Thalassaemia	9	0.77
		HbH Disease	34	2.91
		Others	2	0.17
30 - 34.9	142	Beta Thalassaemia Major	29	2.48
		Beta Thalassaemia Intermedia	61	5.22
		HbE-Beta Thalassaemia	11	0.94
		HbH Disease	36	3.08
		Others	5	0.43

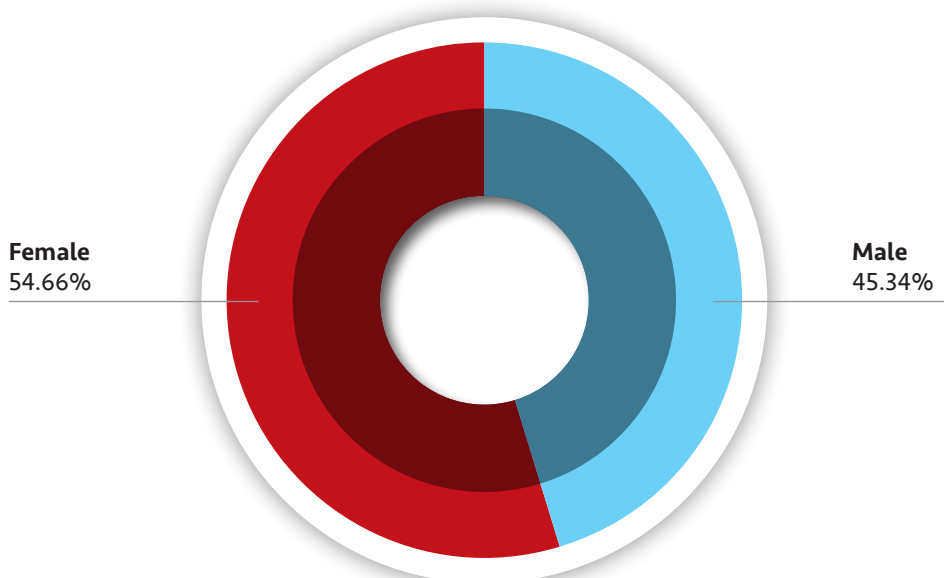
Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	104	Beta Thalassaemia Major	16	1.37
		Beta Thalassaemia Intermedia	48	4.11
		HbE-Beta Thalassaemia	4	0.34
		HbH Disease	25	2.14
		Others	11	0.94
40 - 44.9	84	Beta Thalassaemia Major	4	0.34
		Beta Thalassaemia Intermedia	30	2.57
		HbE-Beta Thalassaemia	9	0.77
		HbH Disease	33	2.82
		Others	8	0.68
45 - 49.9	42	Beta Thalassaemia Major	1	0.09
		Beta Thalassaemia Intermedia	18	1.54
		HbE-Beta Thalassaemia	9	0.77
		HbH Disease	14	1.20
		Others	0	0.00
50 - 54.9	25	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	6	0.51
		HbE-Beta Thalassaemia	4	0.34
		HbH Disease	12	1.03
		Others	3	0.26
55 - 59.9	17	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	5	0.43
		HbE-Beta Thalassaemia	1	0.09
		HbH Disease	7	0.60
		Others	4	0.34
60 - 64.9	17	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	8	0.68
		HbE-Beta Thalassaemia	2	0.17
		HbH Disease	7	0.60
		Others	0	0.00
Above 65	24	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	0.17
		HbE-Beta Thalassaemia	4	0.34
		HbH Disease	17	1.45
		Others	1	0.09
Total			1169	100.00

15 Selangor

15.2.2 Gender

There are more female than male thalassaemia patients in Selangor, at 639 (54.66%) and 530 (45.34%), respectively.

Figure 15.3: Distribution of Patients in Selangor by Gender



252

Table 15.6: Distribution of Patients in Selangor According to Gender by Centre

Centre	Male		Female	
	No.	%	No.	%
Hospital Ampang	280	23.95	423	36.18
Hospital Banting	13	1.11	3	0.26
Hospital Kajang	26	2.22	17	1.45
Hospital Kuala Kubu Bharu	2	0.17	1	0.09
Hospital Selayang	38	3.25	44	3.76
Hospital Serdang	21	1.80	14	1.20
Hospital Sungai Buloh	40	3.42	23	1.97
Hospital Tanjung Karang	0	0.00	3	0.26
HTAJ, Sabak Bernam	3	0.26	4	0.34
HTAR, Klang	101	8.64	100	8.55
Hospital Shah Alam	6	0.51	7	0.60
Total	530	45.34	639	54.66

15.2.3 Ethnic Group

Ethnic groups in Selangor is comprised of Malays (68.60%), Chinese (23.40%), Indian (7.00%) and Others (1.00%). Malay patients form the largest group of patients in Selangor with 902 patients (77.16%), followed by the Chinese with 190 patients (16.25%), patients of other ethnicities with 47 patients (4.02%), Kadazan-Dusun with 16 patients (1.37%), Indian with 7 patients (0.60%), and Orang Asli also with 7 patients (0.60%).

Table 15.7: Distribution of Patients in Selangor by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	902	77.16
Chinese	190	16.25
Indian	7	0.60
Kadazan-Dusun	16	1.37
Orang Asli	7	0.60
Others	47	4.02
Total	1169	100.00

Figure 15.4: Distribution of Patients in Selangor by Ethnic Group

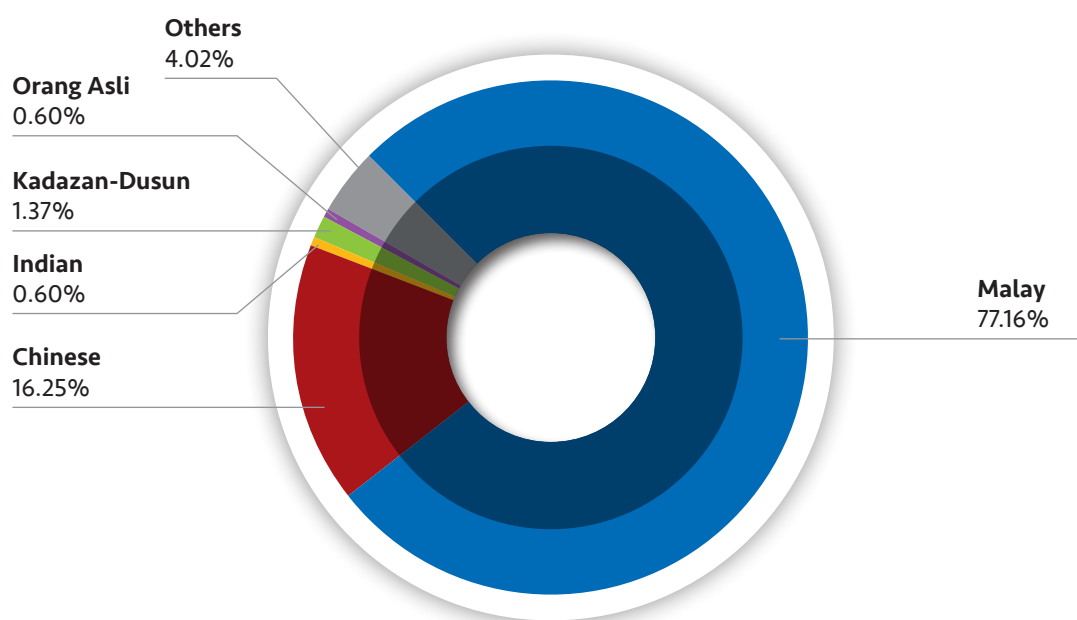


Table 15.8: Distribution of Patients in Selangor According to Ethnic Group by Centre

Centre	Malay		Chinese		Indian		Kadazan		Orang Asli		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Ampang	505	43.20	157	13.43	3	0.26	7	0.60	4	0.34	27	2.31
Hospital Banting	16	1.37	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Kajang	34	2.91	3	0.26	0	0.00	0	0.00	2	0.17	4	0.34
Hospital Kuala Kubu Bharu	2	0.17	1	0.09	0	0.00	0	0.00	0	0.00	0	0.00
Hospital Selayang	60	5.13	17	1.45	1	0.09	1	0.09	0	0.00	3	0.26
Hospital Serdang	30	2.57	1	0.09	0	0.00	3	0.26	0	0.00	1	0.09
Hospital Shah Alam	12	1.03	0	0.00	0	0.00	0	0.00	0	0.00	1	0.09
Hospital Sungai Buloh	58	4.96	2	0.17	0	0.00	0	0.00	0	0.00	3	0.26
Hospital Tanjung Karang	3	0.26	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
HTAJ, Sabak Bernam	7	0.60	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
HTAR, Klang	175	14.97	9	0.77	3	0.26	5	0.43	1	0.09	8	0.68
Total	902	77.16	190	16.25	7	0.60	16	1.37	7	0.60	47	4.02

15.3 Diagnosis

Based on Table 15.9, HbE-beta thalassaemia was diagnosed for 518 of the patients in Selangor (44.31%). This is followed by HbH disease (289 patients, 24.72%), beta thalassaemia major (209 patients, 17.88%), and beta thalassaemia intermedia (89 patients, 7.61%). The remaining 64 patients (5.47%) are being followed up for other forms of haemoglobinopathy.

Table 15.9: Distribution of Patients in Selangor by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	209	17.88
Beta Thalassaemia Intermedia	89	7.61
HbE-Beta Thalassaemia	518	44.31
HbH Disease	289	24.72
Others	64	5.47
Total	1169	100.00

Figure 15.5: Distribution of Patients in Selangor by Diagnosis

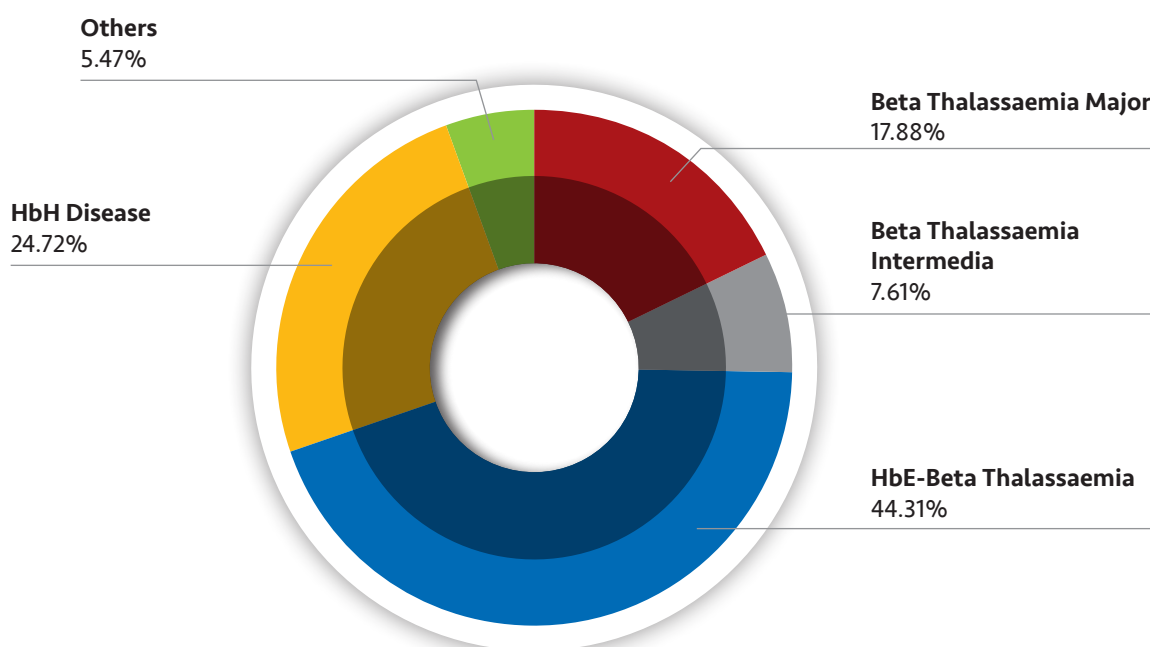


Table 15.10: Distribution of Patients in Selangor According to Diagnosis by Centre

Centre	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others*	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Ampang	121	10.35	42	3.59	287	24.55	213	18.22	40	3.42
Hospital Banting	2	0.17	0	0.0	12	1.03	2	0.17	0	0.00
Hospital Kajang	10	0.86	4	0.34	16	1.37	9	0.77	4	0.34
Hospital Kuala Kubu Bharu	1	0.09	0	0.0	1	0.09	1	0.09	0	0.00
Hospital Selayang	15	1.28	4	0.34	28	2.40	26	2.22	9	0.77
Hospital Serdang	6	0.51	4	0.34	23	1.97	1	0.09	1	0.09
Hospital Shah Alam	2	0.17	0	0.0	8	0.68	3	0.26	0	0.00
Hospital Sungai Buloh	13	1.11	4	0.34	33	2.82	9	0.77	4	0.34
Hospital Tanjung Karang	0	0.0	0	0.0	3	0.26	0	0.00	0	0.00
HTAJ,Sabak Bernam	0	0.0	0	0.0	7	0.60	0	0.00	0	0.00
HTAR,Klang	39	3.34	31	2.65	100	8.55	25	2.14	6	0.51
Total	209	17.88	89	7.61	518	44.31	289	24.72	64	5.47

*Other diagnoses includes alpha thalassaemia.

Table 15.11: Distribution of Patients in Selangor According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	209	Malay	111	9.50
		Chinese	68	5.82
		Indian	3	0.26
		Kadazan-Dusun	12	1.03
		Others	15	1.28
Beta Thalassaemia Intermedia	89	Malay	69	5.90
		Chinese	15	1.28
		Indian	1	0.09
		Kadazan-Dusun	1	0.09
		Others	3	0.26
HbE-Beta Thalassaemia	518	Malay	473	40.46
		Chinese	18	1.54
		Indian	1	0.09
		Kadazan-Dusun	2	0.17
		Orang Asli	3	0.26
		Others	21	1.80
HbH Disease	289	Malay	201	17.19
		Chinese	77	6.59
		Indian	1	0.09
		Kadazan-Dusun	1	0.09
		Orang Asli	3	0.26
		Others	6	0.51
Others	64	Malay	48	4.11
		Chinese	12	1.03
		Indian	1	0.09
		Orang Asli	1	0.09
		Others	2	0.17
Total			1169	100.00

15.4 Treatment

15.4.1 Iron Chelation Therapy

Based Table 15.12, 753 patients in Selangor receive iron chelation therapy. Of these, 292 patients (38.78%) are on oral DFP monotherapy, 161 patients (21.38%) are on DFX monotherapy, and 75 patients (9.96%) patients are on DFO monotherapy. A total of 198 patients (26.29%) are on DFO/DFP combination therapy, 18 patients (2.39%) are on DFO/DFX combination therapy, and 8 patients (1.06%) are on DFP/DFX combination therapy. Only 1 patient (0.13%) receive all three types of chelators (DFO/DFP/DFX).

Table 15.12: Distribution of Patients in Selangor by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	75	9.96
DFP only	292	38.78
DFX only	161	21.38
DFO + DFP	198	26.29
DFP + DFX	8	1.06
DFO + DFX	18	2.39
DFO + DFP + DFX	1	0.13
Total	753	100.00

As shown in Table 15.13, DFX is the most common iron chelator used in Hospital Selayang, Hospital Serdang and Hospital Shah Alam, as thalassaemia patients in these hospitals are of the paediatrics age group. On the other hand, Hospital Banting and Hospital Kajang uses DFO most frequently.

Table 15.13: Distribution of Patients in Selangor According to Type of Iron Chelator Received by Centre

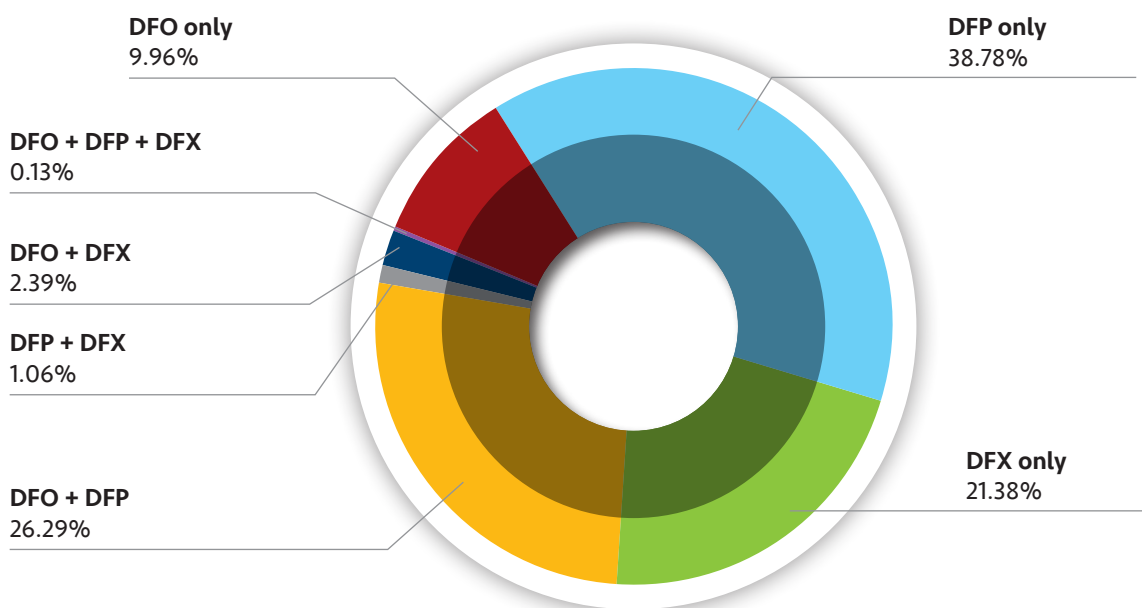
Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Ampang	456	DFO only	31	4.12
		DFP only	241	32.01
		DFX only	20	2.66
		DFO + DFP	146	19.39
		DFP + DFX	7	0.93
		DFO + DFX	10	1.33
		DFO + DFP + DFX	1	0.13
Banting	13	DFO only	6	0.80
		DFP only	2	0.27
		DFX only	3	0.40
		DFO + DFP	2	0.27
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

15 Selangor

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Kajang	22	DFO only	7	0.93
		DFP only	0	0.00
		DFX only	5	0.66
		DFO + DFP	10	1.33
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Kuala Kubu Bharu	3	DFO only	1	0.13
		DFP only	1	0.13
		DFX only	0	0.00
		DFO + DFP	1	0.13
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Selayang	37	DFO only	5	0.66
		DFP only	0	0.00
		DFX only	28	3.72
		DFO + DFP	0	0.00
		DFP + DFX	1	0.13
		DFO + DFX	3	0.40
		DFO + DFP + DFX	0	0.00
Serdang	25	DFO only	6	0.80
		DFP only	0	0.00
		DFX only	18	2.39
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.13
		DFO + DFP + DFX	0	0.00
Shah Alam	4	DFO only	1	0.13
		DFP only	0	0.00
		DFX only	3	0.40
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Sungai Buloh	42	DFO only	3	0.40
		DFP only	17	2.26
		DFX only	9	1.20
		DFO + DFP	13	1.73
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
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
Sabak Bernam	5	DFO only	2	0.27
		DFP only	0	0.00
		DFX only	1	0.13
		DFO + DFP	2	0.27
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Klang	146	DFO only	13	1.73
		DFP only	31	4.12
		DFX only	74	9.83
		DFO + DFP	24	3.19
		DFP + DFX	0	0.00
		DFO + DFX	4	0.53
		DFO + DFP + DFX	0	0.00
Total			753	100.00

Figure 15.6: Distribution of Patients in Selangor by Type of Iron Chelator Received



The type of iron chelator received by age group is shown in Table 15.14. For patients in the age group of 20 years old and above, DFP is the most common iron chelator used.

Table 15.14: Distribution of Patients in Selangor According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	23	DFO only	1	0.13
		DFP only	0	0.00
		DFX only	21	2.79
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	1	0.13
		DFO + DFP + DFX	0	0.00
5-9.9	74	DFO only	11	1.46
		DFP only	11	1.46
		DFX only	48	6.37
		DFO + DFP	2	0.27
		DFP + DFX	1	0.13
		DFO + DFX	1	0.13
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
10-14.9	91	DFO only	13	1.73
		DFP only	12	1.59
		DFX only	47	6.24
		DFO + DFP	15	1.99
		DFP + DFX	1	0.13
		DFO + DFX	3	0.40
		DFO + DFP + DFX	0	0.00
15-19.9	82	DFO only	10	1.33
		DFP only	18	2.39
		DFX only	26	3.45
		DFO + DFP	21	2.79
		DFP + DFX	3	0.40
		DFO + DFX	3	0.40
		DFO + DFP + DFX	1	0.13
20-24.9	121	DFO only	9	1.20
		DFP only	45	5.98
		DFX only	8	1.06
		DFO + DFP	53	7.04
		DFP + DFX	1	0.13
		DFO + DFX	5	0.66
		DFO + DFP + DFX	0	0.00
25-29.9	90	DFO only	14	1.86
		DFP only	41	5.44
		DFX only	1	0.13
		DFO + DFP	32	4.25
		DFP + DFX	1	0.13
		DFO + DFX	1	0.13
		DFO + DFP + DFX	0	0.00
30-34.9	90	DFO only	10	1.33
		DFP only	42	5.58
		DFX only	0	0.00
		DFO + DFP	35	4.65
		DFP + DFX	1	0.13
		DFO + DFX	2	0.27
		DFO + DFP + DFX	0	0.00

15 Selangor

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
35-39.9	60	DFO only	2	0.27
		DFP only	38	5.05
		DFX only	1	0.13
		DFO + DFP	18	2.39
		DFP + DFX	0	0.00
		DFO + DFX	1	0.13
		DFO + DFP + DFX	0	0.00
40-44.9	49	DFO only	1	0.13
		DFP only	36	4.78
		DFX only	1	0.13
		DFO + DFP	10	1.33
		DFP + DFX	0	0.00
		DFO + DFX	1	0.13
		DFO + DFP + DFX	0	0.00
45-49.9	31	DFO only	3	0.40
		DFP only	20	2.66
		DFX only	2	0.27
		DFO + DFP	6	0.80
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50-54.9	13	DFO only	1	0.13
		DFP only	6	0.80
		DFX only	2	0.27
		DFO + DFP	4	0.53
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	7	DFO only	0	0.00
		DFP only	6	0.80
		DFX only	0	0.00
		DFO + DFP	1	0.13
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
60-64.9	11	DFO only	0	0.00
		DFP only	9	1.20
		DFX only	1	0.13
		DFO + DFP	1	0.13
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	11	DFO only	0	0.00
		DFP only	8	1.06
		DFX only	3	0.40
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			753	100.00

15.4.2 Serum Ferritin Level

A total of 492 TDT patients have had their serum ferritin levels measured in 2018. 63 patients (12.80%) have serum ferritin level of lower than 1000 ng/mL. There are 178 patients (36.18%) with serum ferritin level between 1000-2499 ng/mL. However, 109 patients (22.15%) still have a serum ferritin level higher than 5000 ng/mL. Most of the patients with high serum ferritin levels are adolescents and adults who are likely to be non-compliant to iron chelating agents.

Table 15.15: Distribution of Patients in Selangor According to Most Recent Serum Ferritin Level by Centre in 2018

Serum Ferritin Level (ng/mL)	Centre	Total	< 1000		1000-2499		2500-4999		5000-9999		10,000+	
			No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Ampang	265	38	7.72	82	16.67	64	13.01	61	12.40	20	4.07	
Hospital Banting	13	2	0.41	5	1.02	5	1.02	1	0.20	0	0.00	
Hospital Kajang	17	2	0.41	9	1.83	4	0.81	2	0.41	0	0.00	
Hospital Kuala Kubu Baru	1	0	0.00	0	0.00	1	0.20	0	0.00	0	0.00	
Hospital Selayang	28	5	1.02	12	2.44	9	1.83	2	0.41	0	0.00	
Hospital Serdang	25	2	0.41	7	1.42	12	2.44	2	0.41	2	0.41	
Hospital Shah Alam	7	3	0.61	4	0.81	0	0.00	0	0.00	0	0.00	
Hospital Sungai Buloh	42	3	0.61	19	3.86	16	3.25	4	0.81	0	0.00	
Hospital Tanjung Karang	0	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	
Hospital Sabak Bernam	1	0	0.00	1	0.20	0	0.00	0	0.00	0	0.00	
HTAR,Klang	93	8	1.63	39	7.93	31	6.30	12	2.44	3	0.61	
Total	492	63	12.80	178	36.18	142	28.86	84	17.07	25	5.08	

15.5 Observation and Comment

Male to female patients ratio in Selangor was near 1:1. By ethnicity, Malay patients form the largest number of patients in Selangor. Patients age group of 20 to 24.9 years has the highest number of patients. Most adult patients are treated in Hospital Ampang, as it is the country’s referral haematology centre. One third of patients in Selangor are in the paediatric age group, indicating poor awareness on thalassaemia inheritance.

Most patients were diagnosed with HbE-beta thalassaemia. Other haemoglobinopathies include beta thalassaemia major, HbH disease, beta thalassaemia intermedia and more. All these diagnoses were based on Hb and DNA analysis for identification of specific mutations.

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

Many TDT patients in Selangor have serum ferritin level between 1000-2500 ng/mL. Most NTD patients’ serum ferritin levels are not regularly monitored and their appointments are at longer intervals compared to TDT patients. Furthermore, there was a period of 3-4 months whereby the test was not offered in the hospital due to budget constraints. Although high ferritin level represents iron overload, this positive acute phase proteins may be increased during acute and chronic disorders. Therefore, the health status of thalassaemia patients may also influence the serum ferritin level.

As a conclusion, in order to prevent thalassaemia from becoming a disease burden to the country, more thalassaemia screening and public education regarding thalassaemia should be emphasised through various mediums, such as campaigns and social media. Thus, awareness on this genetic disease will bridge the knowledge to the society.

15.5.1 Recommendations

1. To have clear definition for diagnosis should be implemented and standardised among all centres throughout the country.
2. To have clear and standardised definition of “lost to follow-up” status, i.e. either one year, two years, etc.
3. To clearly define TDT and NTD and subsequently include it in the data that needs to be filled up in the registry.
4. More frequent visits to peripheral hospitals to better update data.

16 Terengganu

16.1 Introduction

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

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

The MTR in Terengganu is currently being coordinated by Dr Nazzlin Dizana Din, SN Norashikin Mustaffa and Mohd Saharudin bin Mat Salim (RA, MTR).

16.2 Patient Demographics

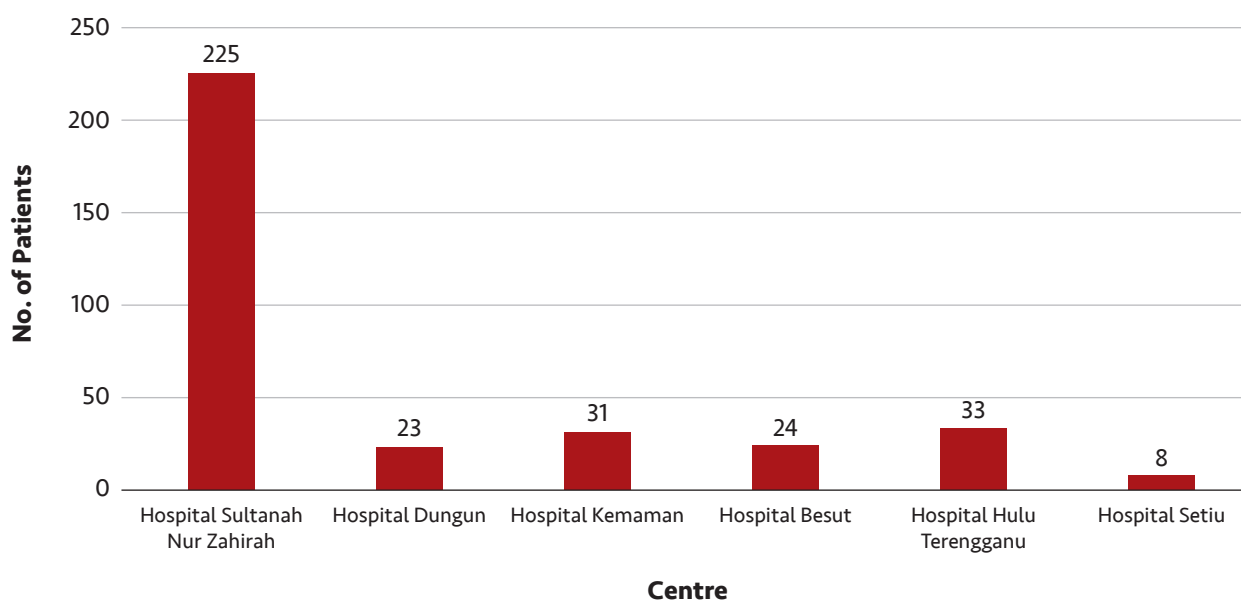
Data analysed were taken from patients with status of either alive, lost to follow-up or cured by transplant. The total number of thalassaemia patients in Terengganu in 2018 is 344 patients, compared to 301 patients in 2017. This reflects an increase of 43 patients (14.29%) in 2018 from 2017.

Table 16.1: Distribution of Patients in Terengganu by Centre

Centre	No. of Patients (n)	Percentage (%)
Hospital Sultanah Nur Zahirah	225	65.41
Hospital Dungun	23	6.69
Hospital Kemaman	31	9.01
Hospital Besut	24	6.98
Hospital Hulu Terengganu	33	9.59
Hospital Setiu	8	2.33
Total	344	100.00

16 Terengganu

Figure 16.1: Distribution of Patients in Terengganu by Centre



266

Table 16.2: Distribution of Patients in Terengganu by Vital Status

Vital Status	Patients
Alive	306
Cumulative Reported Cured by Stem Cell Therapy	1
Lost to Follow-up	37
Total	344
Deaths in 2018	1
Cumulative Reported Deaths	23
Newly Diagnosed in 2018	5

Up to November 2018, there was only one death of thalassaemia patient recorded in this year. One patient who underwent haematopoietic stem cell transplantation (HSCT) in October 2017 has been cured. There were five new cases diagnosed in 2018: one patient each from Hospital Kemaman and Hospital Hulu Terengganu, and the remaining three from HSNZ, Kuala Terengganu.

Table 16.3: Cumulative Causes of Death in Terengganu

Causes of Death	No. of Patients
Infections	9
Cardiac	6
Liver Disease	1
Unknown	7
Total	23

16.2.1 Age

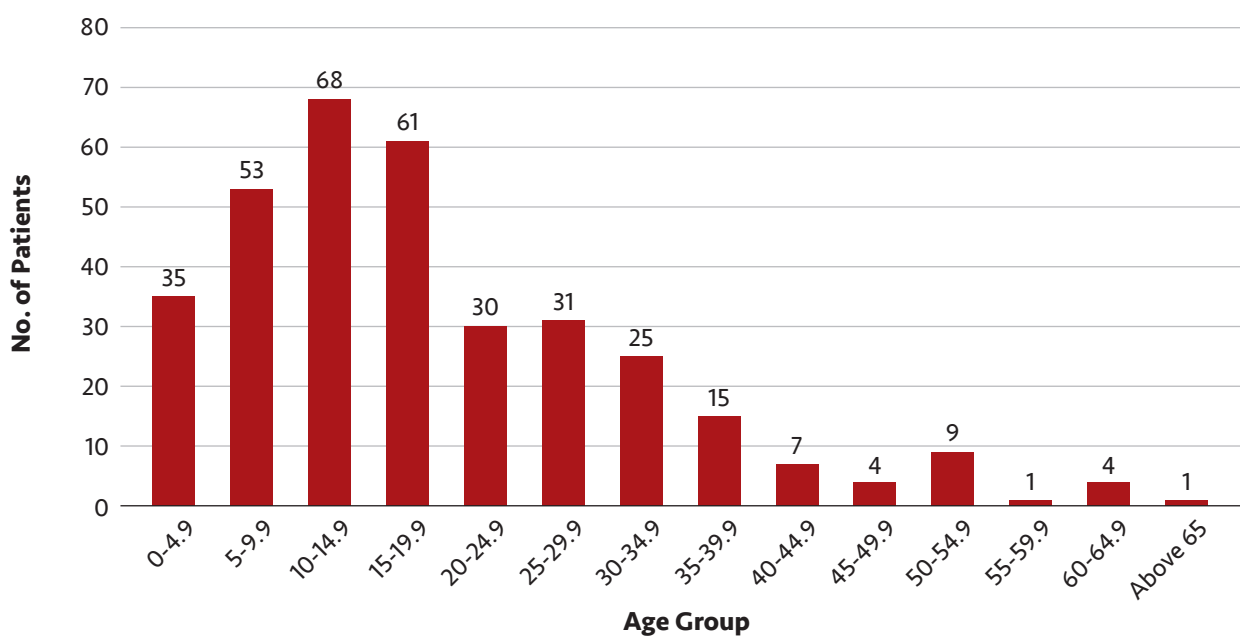
Majority of thalassaemia patients in Terengganu are between the age of 5-19.9 years old (182 of 344 patients, 52.91%).

Table 16.4: Distribution of Patients in Terengganu by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 – 4.9	35	10.17
5 – 9.9	53	15.41
10 – 14.9	68	19.77
15 – 19.9	61	17.73
20 – 24.9	30	8.72
25 – 29.9	31	9.01
30 – 34.9	25	7.27
35 – 39.9	15	4.36
40 – 44.9	7	2.03
45 – 49.9	4	1.16
50 – 54.9	9	2.62
55 – 59.9	1	0.29
60 – 64.9	4	1.16
Above 65	1	0.29
Total	344	100.0

Majority of the patients are managed by the Paediatric Haemato-Oncologist based in HSNZ.

Figure 16.2: Distribution of Patients in Terengganu by Age Group



16 Terengganu

Table 16.5: Distribution of Patients in Terengganu According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	35	Beta Thalassaemia Major	8	2.33
		Beta Thalassaemia Intermedia	1	0.29
		HbE-Beta Thalassaemia	19	5.52
		HbH Disease	6	1.74
		Others	1	0.29
5 - 9.9	53	Beta Thalassaemia Major	4	1.16
		Beta Thalassaemia Intermedia	3	0.87
		HbE-Beta Thalassaemia	32	9.30
		HbH Disease	11	3.20
		Others	3	0.87
10 -14.9	68	Beta Thalassaemia Major	16	4.65
		Beta Thalassaemia Intermedia	3	0.87
		HbE-Beta Thalassaemia	31	9.01
		HbH Disease	16	4.65
		Others	2	0.58
15 -19.9	61	Beta Thalassaemia Major	9	2.62
		Beta Thalassaemia Intermedia	3	0.87
		HbE-Beta Thalassaemia	36	10.47
		HbH Disease	12	3.49
		Others	1	0.29
20 - 24.9	30	Beta Thalassaemia Major	7	2.03
		Beta Thalassaemia Intermedia	1	0.29
		HbE-Beta Thalassaemia	17	4.94
		HbH Disease	4	1.16
		Others	1	0.29
25 - 29.9	31	Beta Thalassaemia Major	9	2.62
		Beta Thalassaemia Intermedia	4	1.16
		HbE-Beta Thalassaemia	16	4.65
		HbH Disease	2	0.58
		Others	0	0.00
30 - 34.9	25	Beta Thalassaemia Major	5	1.45
		Beta Thalassaemia Intermedia	4	1.16
		HbE-Beta Thalassaemia	13	3.78
		HbH Disease	2	0.58
		Others	1	0.29

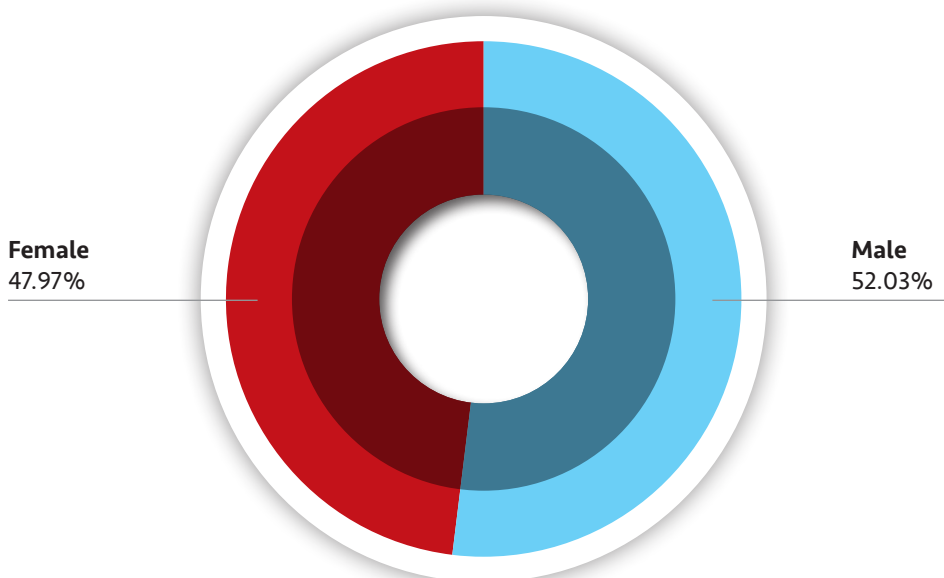
Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	15	Beta Thalassaemia Major	1	0.29
		Beta Thalassaemia Intermedia	3	0.87
		HbE-Beta Thalassaemia	11	3.20
		HbH Disease	0	0.00
		Others	0	0.00
40 - 44.9	7	Beta Thalassaemia Major	1	0.29
		Beta Thalassaemia Intermedia	2	0.58
		HbE-Beta Thalassaemia	2	0.58
		HbH Disease	2	0.58
		Others	0	0.00
45 - 49.9	4	Beta Thalassaemia Major	1	0.29
		Beta Thalassaemia Intermedia	2	0.58
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.29
		Others	0	0.00
50 - 54.9	9	Beta Thalassaemia Major	1	0.29
		Beta Thalassaemia Intermedia	4	1.16
		HbE-Beta Thalassaemia	2	0.58
		HbH Disease	2	0.58
		Others	0	0.00
55 - 59.9	1	Beta Thalassaemia Major	1	0.29
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
60 - 64.9	4	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.29
		HbE-Beta Thalassaemia	1	0.29
		HbH Disease	2	0.58
		Others	0	0.00
Above 65	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	0.29
		Others	0	0.00
Total			344	100.0

16 Terengganu

16.2.2 Gender

There are 165 female patients (47.97%) and 179 male patients (52.03%) in Terengganu. The distribution of the male and female patients in each district hospital is shown in Figure 16.3.

Figure 16.3: Distribution of Patients in Terengganu by Gender



270

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

Centre	Male		Female	
	No.	%	No.	%
HSNZ, Kuala Terengganu	116	33.72	109	31.69
Hospital Dungun	12	3.49	11	3.20
Hospital Kemaman	23	6.69	8	2.33
Hospital Besut	11	3.20	13	3.78
Hospital Hulu Terengganu	16	4.65	17	4.94
Hospital Setiu	1	0.29	7	2.03
Total	179	52.03	165	47.97

16.2.3 Ethnic Group

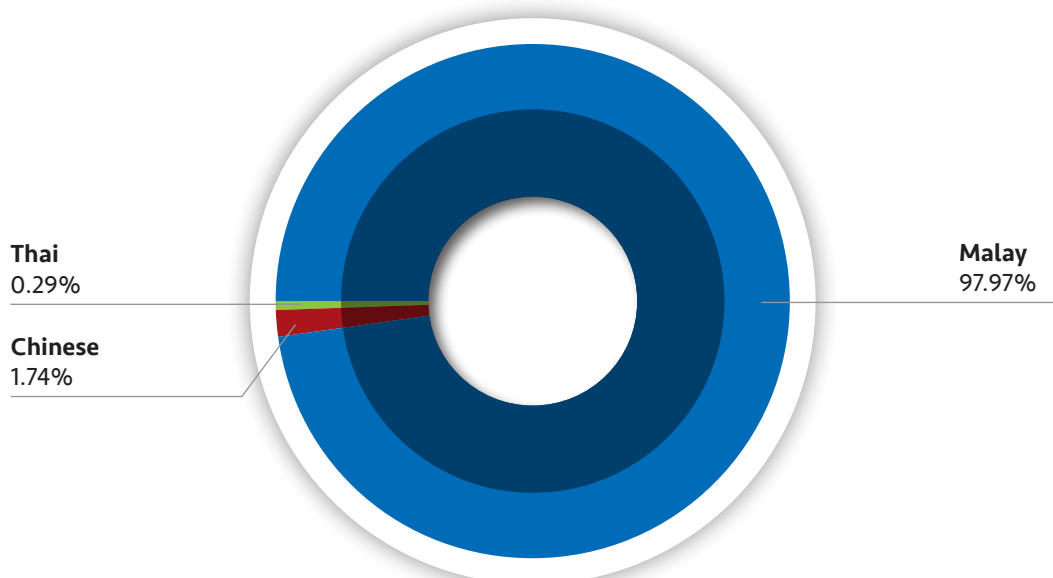
Table 16.7 showed the ethnic distribution of thalassaemia patients in Terengganu. Malay patients form the great majority of patients in the state (337 patients, 97.97%).

Table 16.7: Distribution of Patients in Terengganu by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	337	97.97
Chinese	6	1.74
Others:		
Thai	1	0.29
Total	344	100.00

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

Centre	Malay		Chinese		Thai	
	No.	%	No.	%	No.	%
Hospital SNZ	220	63.95	5	1.45	0	0.00
Hospital Dungun	22	6.40	0	0.00	1	0.29
Hospital Kemaman	30	8.72	1	0.29	0	0.00
Hospital Besut	24	6.98	0	0.00	0	0.00
Hospital Hulu Terengganu	33	9.59	0	0.00	0	0.00
Hospital Setiu	8	2.33	0	0.00	0	0.00
Total	337	97.97	6	1.74	1	0.29

Figure 16.4: Distribution of Patients in Terengganu by Ethnic Group

271

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

16.3 Diagnosis

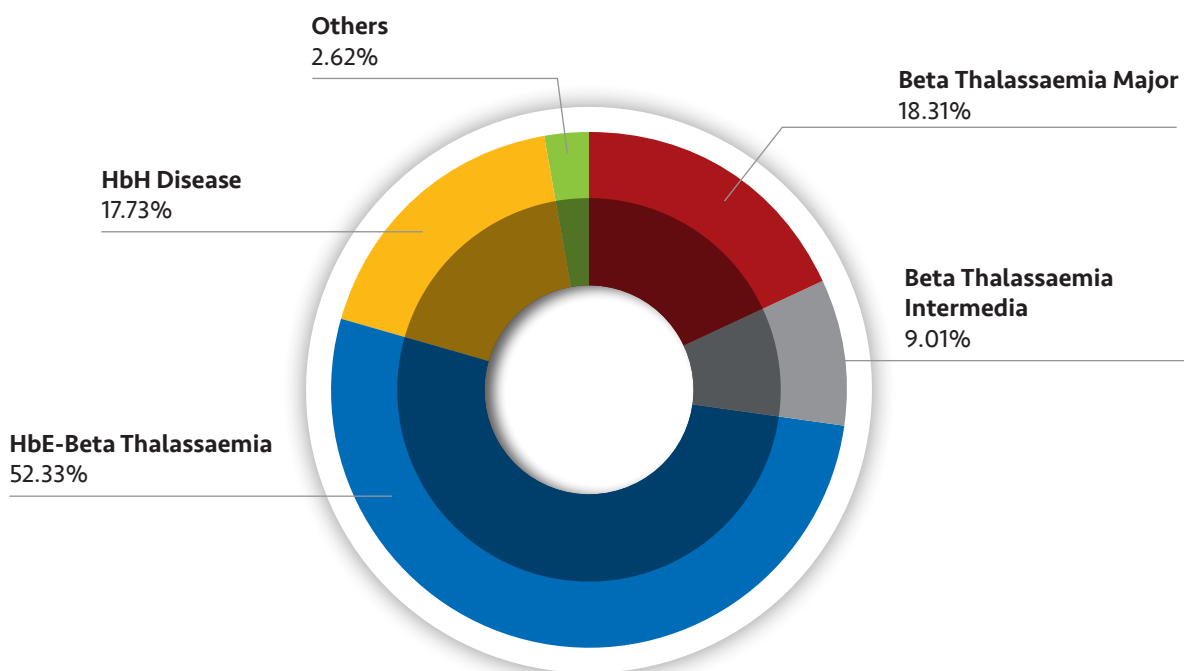
HbE-beta thalassaemia formed the major number of the patients (180 patients, 52.33%), followed by beta thalassaemia major (63 patients, 18.31%) and HbH disease (61 patients, 17.73%).

Table 16.9: Distribution of Patients in Terengganu by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	63	18.31
Beta Thalassaemia Intermedia	31	9.01
HbE-Beta Thalassaemia	180	52.33
HbH Disease	61	17.73
Others	9	2.62
Total	344	100.00

16 Terengganu

Figure 16.5: Distribution of Patients in Terengganu by Diagnosis



272

Table 16.10: Distribution of Patients in Terengganu According to Diagnosis by Centre

Diagnosis	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others	
	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital SNZ	27	7.85	21	6.10	124	36.05	46	13.37	7	2.03
Hospital Dungun	6	1.74	1	0.29	14	4.07	1	0.29	1	0.29
Hospital Kemaman	5	1.45	4	1.16	18	5.23	4	1.16	0	0.00
Hospital Besut	8	2.33	2	0.58	10	2.91	4	1.16	0	0.00
Hospital H.Terengganu	16	4.65	3	0.87	8	2.33	5	1.45	1	0.29
Hospital Setiu	1	0.29	0	0.00	6	1.74	1	0.29	0	0.00
Total	63	18.31	31	9.01	180	52.33	61	17.73	9	2.62

Table 16.11: Distribution of Patients in Terengganu According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	63	Malay	61	17.73
		Chinese	1	0.29
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	1	0.29
Beta Thalassaemia Intermedia	31	Malay	30	8.72
		Chinese	1	0.29
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
HbE-Beta Thalassaemia	180	Malay	179	52.03
		Chinese	1	0.29
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
HbH Disease	61	Malay	58	16.86
		Chinese	3	0.87
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Others	9	Malay	9	2.62
		Chinese	0	0.00
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Total			344	100.00

16 Terengganu

16.4 Treatment

16.4.1 Iron Chelation Therapy

A total of 240 thalassaemia patients (69.77%) in Terengganu has been put on chelating agents. DFP is the commonly prescribed agent (prescribed to 111 patients out of 240, 46.25%), followed by DFX (78 patients, 32.50%) and DFO (29 patients, 12.08%).

Table 16.12: Distribution of Patients in Terengganu by Type of Iron Chelator Received

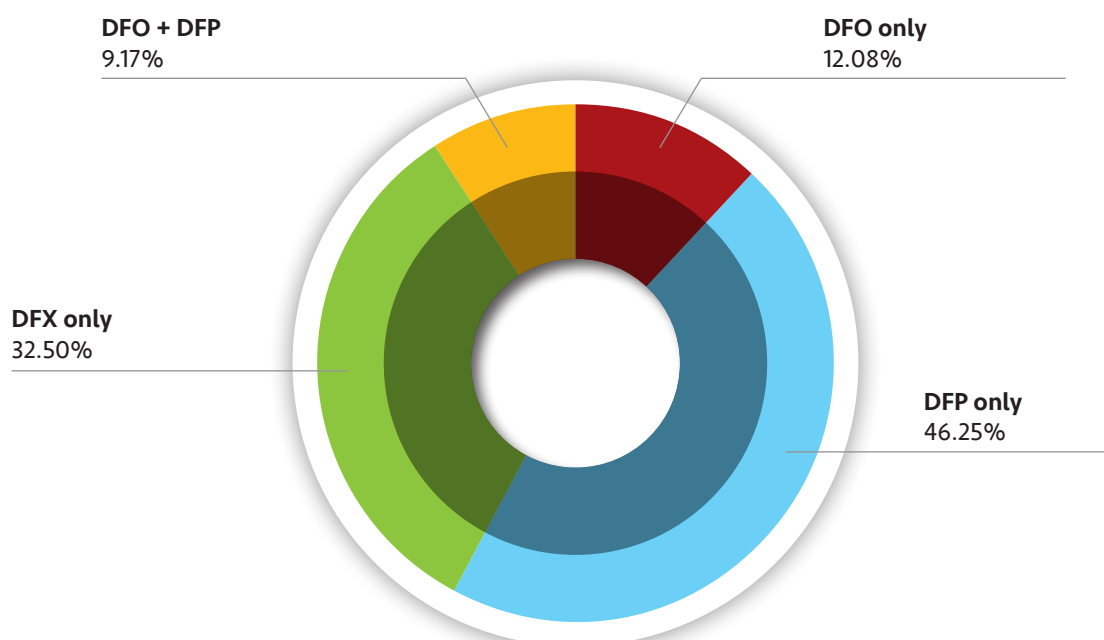
Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	29	12.08
DFP only	111	46.25
DFX only	78	32.50
DFO + DFP	22	9.17
DFP + DFX	0	0.00
DFO + DFX	0	0.00
DFO + DFP + DFX	0	0.00
Total	240	100.00

Table 16.13: Distribution of Patients in Terengganu According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Sultanah Nur Zahirah (HSNZ)	149	DFO only	22	9.17
		DFP only	61	25.42
		DFX only	47	19.58
		DFO + DFP	19	7.92
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Dungun	21	DFO only	1	0.42
		DFP only	13	5.42
		DFX only	6	2.50
		DFO + DFP	1	0.42
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Kemaman	25	DFO only	2	0.83
		DFP only	8	3.33
		DFX only	15	6.25
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Hospital Besut	14	DFO only	2	0.83
		DFP only	6	2.50
		DFX only	6	2.50
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Hulu Terengganu	24	DFO only	2	0.83
		DFP only	17	7.08
		DFX only	3	1.25
		DFO + DFP	2	0.83
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Hospital Setiu	7	DFO only	0	0.00
		DFP only	6	2.50
		DFX only	1	0.42
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			276	100.00

Figure 16.6 : Distribution of Patients in Terengganu by Type of Iron Chelator Received



16 Terengganu

Table 16.14: Distribution of Patients in Terengganu According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	18	DFO only	1	0.42
		DFP only	0	0.00
		DFX only	17	7.08
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5-9.9	36	DFO only	2	0.83
		DFP only	5	2.08
		DFX only	29	12.08
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
10-14.9	55	DFO only	8	3.33
		DFP only	24	10.00
		DFX only	22	9.17
		DFO + DFP	1	0.42
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
15-19.9	42	DFO only	5	2.08
		DFP only	24	10.00
		DFX only	3	1.25
		DFO + DFP	10	4.17
		DFP + DFX	0	0.00
		DFO + DFX	0	0
		DFO + DFP + DFX	0	0
20-24.9	20	DFO only	6	2.50
		DFP only	11	4.58
		DFX only	1	0.42
		DFO + DFP	2	0.83
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
25-29.9	25	DFO only	5	2.08
		DFP only	17	7.08
		DFX only	0	0.00
		DFO + DFP	3	1.25
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30-34.9	15	DFO only	0	0.00
		DFP only	12	5.00
		DFX only	0	0.00
		DFO + DFP	3	1.25
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
35-39.9	12	DFO only	1	0.42
		DFP only	5	2.08
		DFX only	3	1.25
		DFO + DFP	3	1.25
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40-44.9	4	DFO only	1	0.42
		DFP only	2	0.83
		DFX only	1	0.42
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-49.9	2	DFO only	0	0.00
		DFP only	2	0.83
		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

16 Terengganu

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
50-54.9	7	DFO only	0	0.00
		DFP only	7	2.92
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60-64.9	3	DFO only	0	0.00
		DFP only	1	0.42
		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
Above 65	1	DFO only	0	0.00
		DFP only	1	0.42
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			240	100.00

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

16.4.2 Serum Ferritin Level

There were 221 (90.20%) out of 245 TDT patients who had their serum ferritin levels recorded and updated in their notes. About 135 patients (55.10%) of the 245 TDT patients has a serum ferritin level lower than 2500 ng/mL. The remaining 86 patients (35.10%) has a serum ferritin level higher than 2500 ng/mL, including five patients with serum ferritin level exceeding 10000 ng/mL.

Table 16.15: Distribution of Patients in Terengganu According to Most Recent Serum Ferritin Level by Centre

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital SNZ	149	50	22.62	49	22.17	34	15.38	14	6.33	2	0.90
Hospital Dungun	11	2	0.90	6	2.71	2	0.90	1	0.45	0	0.00
Hospital Kemaman	22	4	1.81	8	3.62	6	2.71	4	1.81	0	0.00
Hospital Besut	6	1	0.45	3	1.36	0	0.00	2	0.90	0	0.00
Hospital Hulu Terengganu	28	4	1.81	7	3.17	8	3.62	6	2.71	3	1.36
Hospital Setiu	5	0	0.00	1	0.45	4	1.81	0	0.00	0	0.00
Total	221	61	27.60	74	33.48	54	24.43	27	12.22	5	2.26

16.5 Observation and Comments

The current registry lacks an updated dataset on latest serum ferritin level, intake of iron chelation, complications, etc. This is due to a few factors, particularly pertaining to district hospitals:

- No person-in-charge assigned specifically for handling thalassaemia patients at the district hospitals. Majority of the staff do not understand the importance and needs of the MTR.
- Many staff do not know the purpose of thalassaemia management.
- Poor record management; record of screening and test results were disorganised.

16.5.1 Recommendations

- A standardised chart of thalassaemia and patient diary can be introduced across the state and be made available for every thalassaemia patient. The chart is to be used in continuity. By implementing this step, data can be easily collected by the RA.
- To conduct a regular workshop for healthcare personnel to train them on thalassaemia management.
- Engagement of dedicated healthcare professionals in district hospitals to update patients' data in the MTR.

16.5.2 Conclusions

- Majority of thalassaemia patients in Terengganu receive treatment at tertiary centre, i.e. HSNZ (225 out of 344 patients, 65.41%)
- Patients aged 20 years and below form the majority at 217 patients (63.08%) in Terengganu.
- HbE-beta thalassaemia is the majority diagnosis of the patients in Terengganu (180 patients, 52.33%), followed by beta thalassaemia major (63 patients, 18.31%) and HbH disease (61 patients, 17.73%).
- Nearly all thalassaemia patients in Terengganu are of Malay ethnicity (337 patients, 97.97%).
- Male to female patient ratio is 1:1.
- Based on the current registry data, five new cases were reported in year 2018.
- There was one death of thalassaemia patient in 2018, bringing a cumulative total of 23 deaths recorded in Terengganu.
- One patient underwent HSCT treatment at IPHKL in October 2017. This patient is considered cured.
- DFP is the most prescribed chelating agent, taken by 111 patients (46.25%), followed by DFX.
- DFP/DFO is the most common combination of chelating agents prescribed in Terengganu.
- Patients who are not on chelating agents are mostly of the NTDT type.
- 135 (55.1%) out of 245 TDT patients in Terengganu had an updated serum ferritin level lower than 2500 ng/mL.

Wilayah Persekutuan Kuala Lumpur

17.1 Introduction

The Wilayah Persekutuan consists of three territories, which are Kuala Lumpur, Putrajaya and Labuan. Both Kuala Lumpur and Putrajaya are enclaves in the state of Selangor, while Labuan is an island off coast of the state of Sabah.

Kuala Lumpur is the capital of Malaysia. The city proper, making up an area of 243 km² (94 sq mi), has a population of 1.8 million as of 2018. The ethnic distribution are Malays (40.40%), Chinese (36.43%), Indian (8.41%), and others (14.76%).

There are three major public hospitals in Kuala Lumpur: Institute of Paediatrics, Hospital Kuala Lumpur (IPHKL), Pusat Perubatan Universiti Kebangsaan Malaysia (PPUKM), and Pusat Perubatan Universiti Malaya (PPUM). These three hospitals are the referral centre for paediatric diseases, including those of haematological disorders such as thalassaemia. The centres which perform allogeneic stem cell transplant for thalassaemia patients in Kuala Lumpur are IPHKL, PPUM and Subang Jaya Medical Centre (SJMC).

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

The MTR was started in 2006. Initially, data was collected only for IPHKL and PPUKM using Microsoft Access, after which, the data was migrated to the current website-based registry format. Data entry training for end users commenced in 2007. For Kuala Lumpur, data is collected by temporary RAs for IPHKL, PPUKM and PPUM.

17.2 Patient Demographics

Patients were categorised as alive, lost to follow-up, cured by transplant or deceased. The cumulative number of death of thalassaemia patients in Kuala Lumpur is 79 since 1997. The total number of patients are 535, including 89 patients lost to follow-up and 22 patients cured by transplant.

Table 17.1: Distribution of Patients in Kuala Lumpur by Centre

Centre	Patient Distribution	
	No. of Patients (n)	Percentage (%)
IPHKL	221	41.31
PPUKM	187	34.95
PPUM	127	23.74
Total	535	100.00



Figure 17.1: Distribution of Patients in Kuala Lumpur by Centre

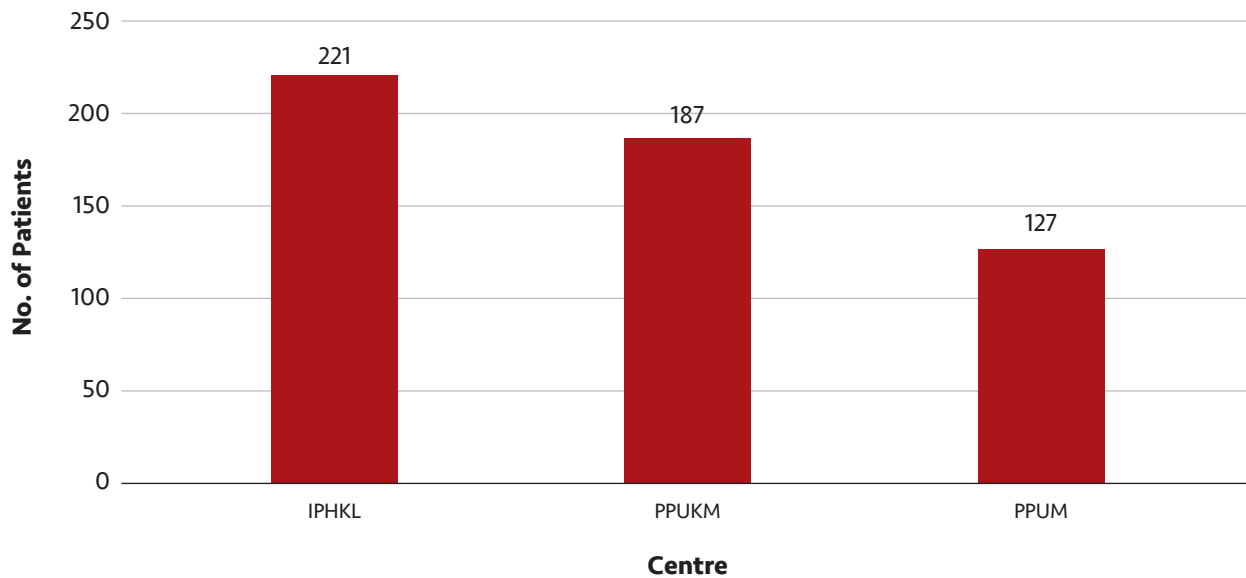


Table 17.2: Distribution of Patients in Kuala Lumpur by Vital Status

Status	Patients
Alive	424
Cumulative Reported Cured by Stem Cell Therapy	22
Lost to Follow-up	89
Total	535
Deaths in 2018	1
Cumulative Reported Deaths	79

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

Cause of Death	No. of Patients
Infections	18
Cardiac Causes	36
Endocrine Complications	1
Others	3
Tumours	1
Unknown	20
Total	79

17.2.1 Age

The youngest patient in Kuala Lumpur is 2 years old and the oldest is 66 years old. Patients aged 10-19.9 years old form the largest group patients in Kuala Lumpur. Of these, a sizeable number is transferred to the adult treatment centre once they reach 18 years of age (based on year of birth).

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

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	46	8.60
5 - 9.9	95	17.76
10 - 14.9	106	19.81
15 - 19.9	104	19.44
20 - 24.9	71	13.27
25 - 29.9	57	10.65
30 - 34.9	40	7.48
35 - 39.9	13	2.43
40 - 44.9	1	0.19
45 - 49.9	0	0.00
50 - 54.9	1	0.19
55 - 59.9	0	0.00
60 - 64.9	0	0.00
Above 65	1	0.19
Total	535	100.00

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

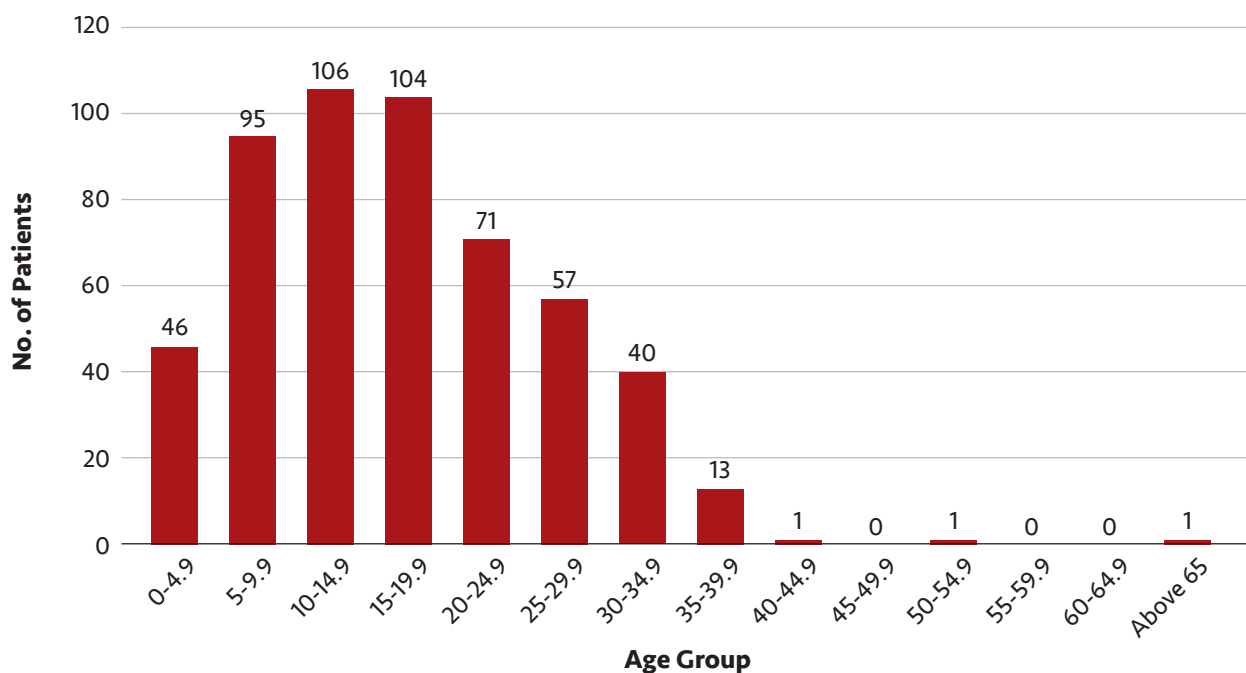


Table 17.5: Distribution of Patients in Kuala Lumpur According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	46	Beta Thalassaemia Major	12	2.24
		Beta Thalassaemia Intermedia	4	0.75
		HbE-Beta Thalassaemia	18	3.36
		HbH Disease	8	1.50
		Others	4	0.75
5 - 9.9	95	Beta Thalassaemia Major	26	4.86
		Beta Thalassaemia Intermedia	4	0.75
		HbE-Beta Thalassaemia	43	8.04
		HbH Disease	15	2.80
		Others	7	1.31
10 -14.9	106	Beta Thalassaemia Major	27	5.05
		Beta Thalassaemia Intermedia	2	0.37
		HbE-Beta Thalassaemia	49	9.16
		HbH Disease	21	3.93
		Others	7	1.31
15 -19.9	104	Beta Thalassaemia Major	27	5.05
		Beta Thalassaemia Intermedia	3	0.56
		HbE-Beta Thalassaemia	51	9.53
		HbH Disease	21	3.93
		Others	2	0.37
20 - 24.9	71	Beta Thalassaemia Major	21	3.93
		Beta Thalassaemia Intermedia	1	0.19
		HbE-Beta Thalassaemia	28	5.23
		HbH Disease	16	2.99
		Others	5	0.93
25 - 29.9	57	Beta Thalassaemia Major	18	3.36
		Beta Thalassaemia Intermedia	2	0.37
		HbE-Beta Thalassaemia	26	4.86
		HbH Disease	11	2.06
		Others	0	0.00
30 - 34.9	40	Beta Thalassaemia Major	16	2.99
		Beta Thalassaemia Intermedia	2	0.37
		HbE-Beta Thalassaemia	15	2.80
		HbH Disease	6	1.12
		Others	1	0.19

17 Wilayah Persekutuan Kuala Lumpur

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	13	Beta Thalassaemia Major	4	0.75
		Beta Thalassaemia Intermedia	1	0.19
		HbE-Beta Thalassaemia	6	1.12
		HbH Disease	2	0.37
		Others	0	0.00
40 - 44.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	1	0.19
		HbH Disease	0	0.00
		Others	0	0.00
45 - 49.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
50 - 54.9	1	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	1	0.19
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
55 - 59.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
60 - 64.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	1	Beta Thalassaemia Major	1	0.19
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			535	100.00

17.2.2 Gender

The gender distribution of thalassaemia patients in Kuala Lumpur are nearly equal. There are 287 male patients (53.64%) and 248 female patients (46.36%) in Kuala Lumpur.

Figure 17.3: Distribution of Patients in Kuala Lumpur by Gender

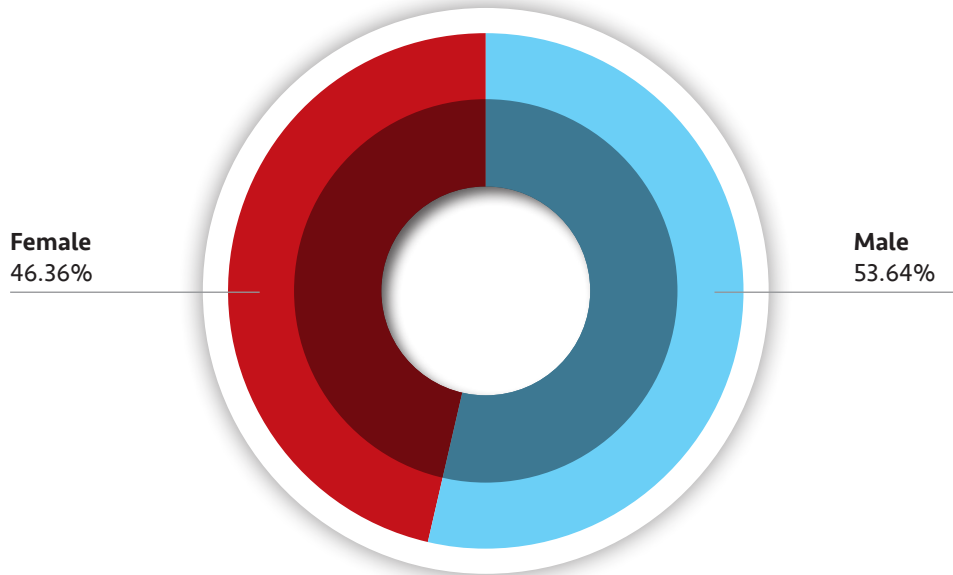


Table 17.6: Distribution of Patients in Kuala Lumpur by Gender

Centre	Male		Female	
	No.	%	No.	%
IPHKL	119	22.24	102	19.07
PPUKM	100	18.69	87	16.26
PPUM	68	12.71	59	11.03
Total	287	53.64	248	46.36

17 Wilayah Persekutuan Kuala Lumpur

17.2.3 Ethnic Group

A total of 393 patients (73.46%) in Kuala Lumpur are Malay, followed by 113 Chinese patients (21.12%) and three Indian patients (0.56%). Eleven non-Malaysians are also seeking treatment in Kuala Lumpur. For the non-citizens, the treatment fees are as per foreigners with self-purchase of iron chelators.

Table 17.7: Distribution of Patients in Kuala Lumpur by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	393	73.46
Chinese	113	21.12
Indian	3	0.56
Kadazan-Dusun	2	0.37
Thai (Malaysian)	2	0.37
Murut	1	0.19
Bidayuh	1	0.19
Mixed	2	0.37
Foreigner	11	2.06
Others	7	1.31
Total	535	100

Figure 17.4: Distribution of Patients in Kuala Lumpur by Ethnic Group

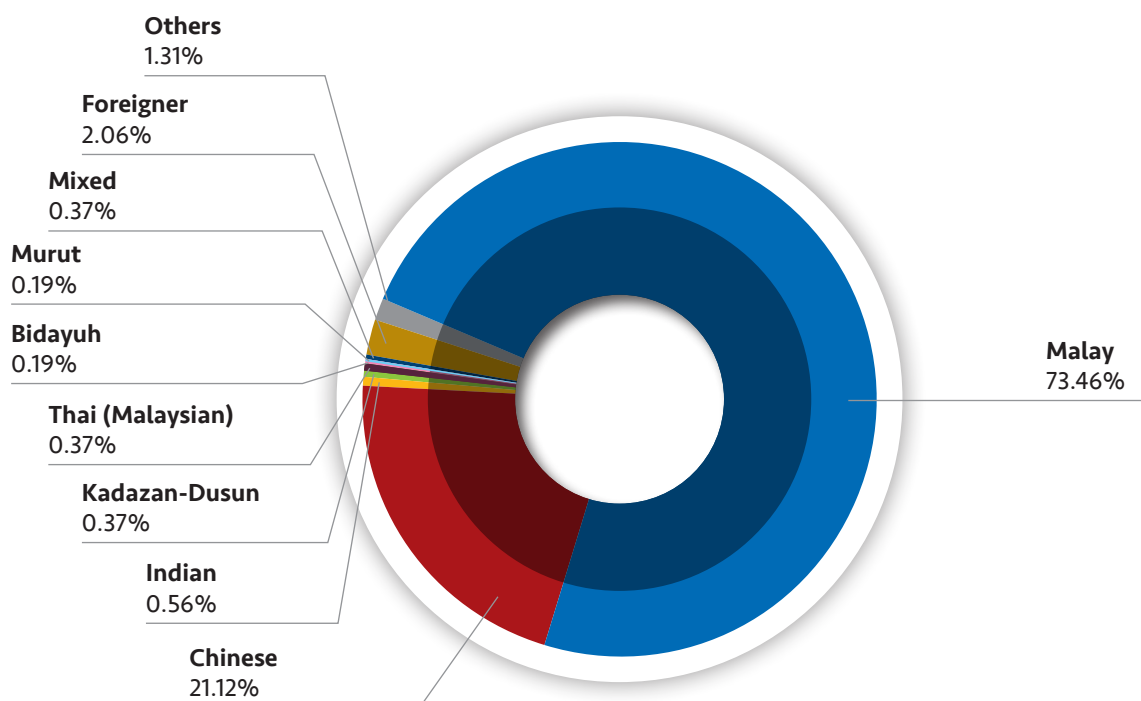


Table 17.8: Distribution of Patients in Kuala Lumpur According to Ethnic Group by Centre

Centre	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
IPHKL	221	Malay	170	31.78
		Chinese	32	5.98
		Indian	1	0.19
		Kadazan-Dusun	2	0.37
		Thai	1	0.19
		Mixed	2	0.37
		Foreigner	8	1.50
		Others	5	0.93
PPUKM	187	Malay	142	26.54
		Chinese	41	7.66
		Murut	1	0.19
		Bidayuh	1	0.19
		Foreigner	2	0.37
PPUM	127	Malay	81	15.14
		Chinese	40	7.48
		Indian	2	0.37
		Thai	1	0.19
		Foreigner	1	0.19
		Others	2	0.37
Total			535	100.00

17.3 Diagnosis

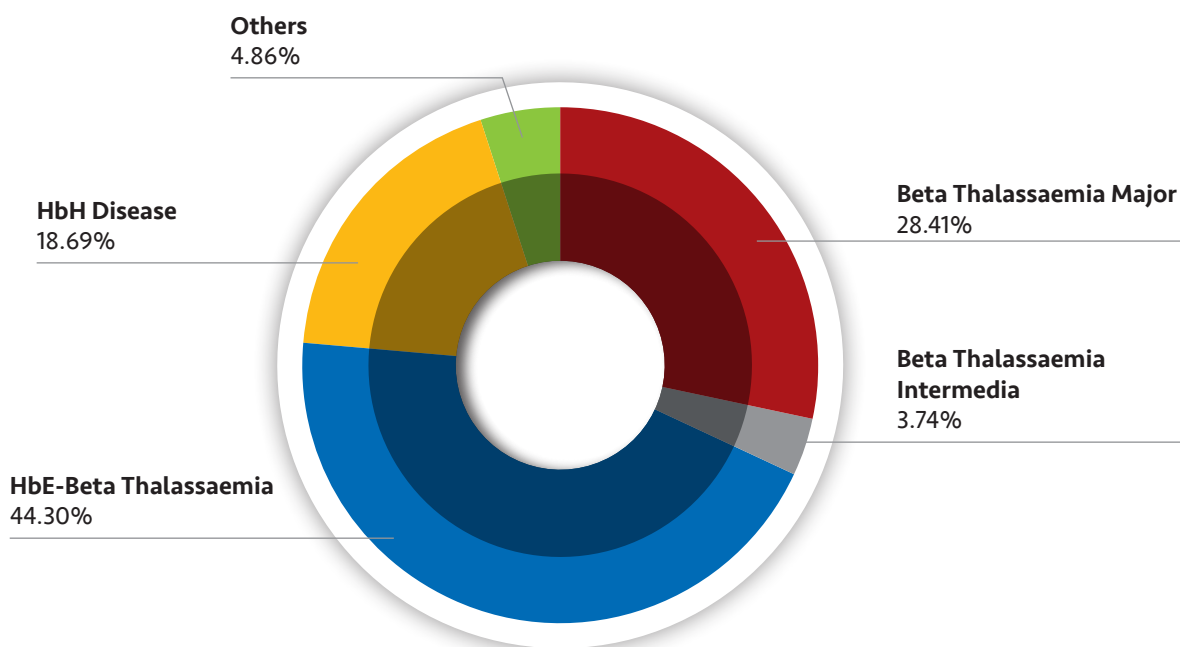
The diagnoses of HbE-beta thalassaemia and beta thalassaemia major are made in 44.30% and 28.41% of the patients in Kuala Lumpur (237 and 152 patients, respectively). Another notable diagnosis is HbH disease with 100 patients diagnosed (18.69%). Twenty-six patients (4.86%) had other diagnosis.

Table 17.9: Distribution of Patients in Kuala Lumpur by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	152	28.41
Beta Thalassaemia Intermedia	20	3.74
HbE-Beta Thalassaemia	237	44.30
HbH Disease	100	18.69
Others	26	4.86
Total	535	100.00

17 Wilayah Persekutuan Kuala Lumpur

Figure 17.5: Distribution of Patients in Kuala Lumpur by Diagnosis



288

MALAYSIAN THALASSAEMIA REGISTRY REPORT 2018

Table 17.10: Distribution of Patients in Kuala Lumpur According to Diagnosis by Centre

Diagnosis	Beta Thalassaemia Major		Beta Thalassaemia Intermedia		HbE-Beta Thalassaemia		HbH Disease		Others*	
	No.	%	No.	%	No.	%	No.	%	No.	%
IPHKL	53	9.91	7	1.31	88	16.45	57	10.65	16	2.99
PPUKM	31	5.79	11	2.06	101	18.88	36	6.73	8	1.50
PPUM	68	12.71	2	0.37	48	8.97	7	1.31	2	0.37
Total	152	28.41	20	3.74	237	44.30	100	18.69	26	4.86

*Includes Hb Adana, delta-beta thalassaemia, HbS, Hb Lepore, etc.

Table 17.11: Distribution of Patients in Kuala Lumpur According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	152	Malay	77	14.39
		Chinese	63	11.78
		Indian	1	0.19
		Kadazan-Dusun	1	0.19
		Others	10	1.87
Beta Thalassaemia Intermedia	20	Malay	12	2.24
		Chinese	5	0.93
		Indian	1	0.19
		Kadazan-Dusun	0	0.00
		Others	2	0.37

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
HbE-Beta Thalassaemia	237	Malay	207	38.69
		Chinese	27	5.05
		Indian	0	0.00
		Kadazan-Dusun	1	0.19
		Others	2	0.37
HbH Disease	100	Malay	73	13.64
		Chinese	27	5.05
		Indian	0	0.00
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Others	26	Malay	24	4.49
		Chinese	1	0.19
		Indian	1	0.19
		Kadazan-Dusun	0	0.00
		Others	0	0.00
Total			535	100.00

17.4 Treatment

17.4.1 Iron Chelation Therapy

Currently, there are 306 TDT and NTD patients (57.20%) on iron chelation therapy in Kuala Lumpur. Patients who are not receiving chelation therapy in Kuala Lumpur number at 229 (42.80%). From the total number of patients receiving iron chelation therapy (306), 237 patients (77.45%) are on monotherapy and 69 patients (22.55%) are on a combination of chelators. The main combination of chelations are DFO and DFP, prescribed to 53 patients (17.32%).

Table 17.12: Distribution of Patients in Kuala Lumpur by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	70	22.88
DFP only	39	12.75
DFX only	128	41.83
DFO + DFP	53	17.32
DFP + DFX	2	0.65
DFO + DFX	14	4.58
DFO + DFP + DFX	0	0.00
Total	306	100.00

Table 17.13: Distribution of Patients in Kuala Lumpur According to Type of Iron Chelator Received by Centre

Centre	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
IPHKL	101	DFO only	11	3.59
		DFP only	0	0.00
		DFX only	75	24.51
		DFO + DFP	3	0.98
		DFP + DFX	0	0.00
		DFO + DFX	12	3.92
		DFO + DFP + DFX	0	0.00
PPUKM	93	DFO only	17	5.56
		DFP only	20	6.54
		DFX only	33	10.78
		DFO + DFP	21	6.86
		DFP + DFX	1	0.33
		DFO + DFX	1	0.33
		DFO + DFP + DFX	0	0.00
PPUM	112	DFO only	42	13.73
		DFP only	19	6.21
		DFX only	20	6.54
		DFO + DFP	29	9.48
		DFP + DFX	1	0.33
		DFO + DFX	1	0.33
		DFO + DFP + DFX	0	0.00
Total			306	100.00

Figure 17.6: Distribution of Patients in Kuala Lumpur by Type of Iron Chelator Received

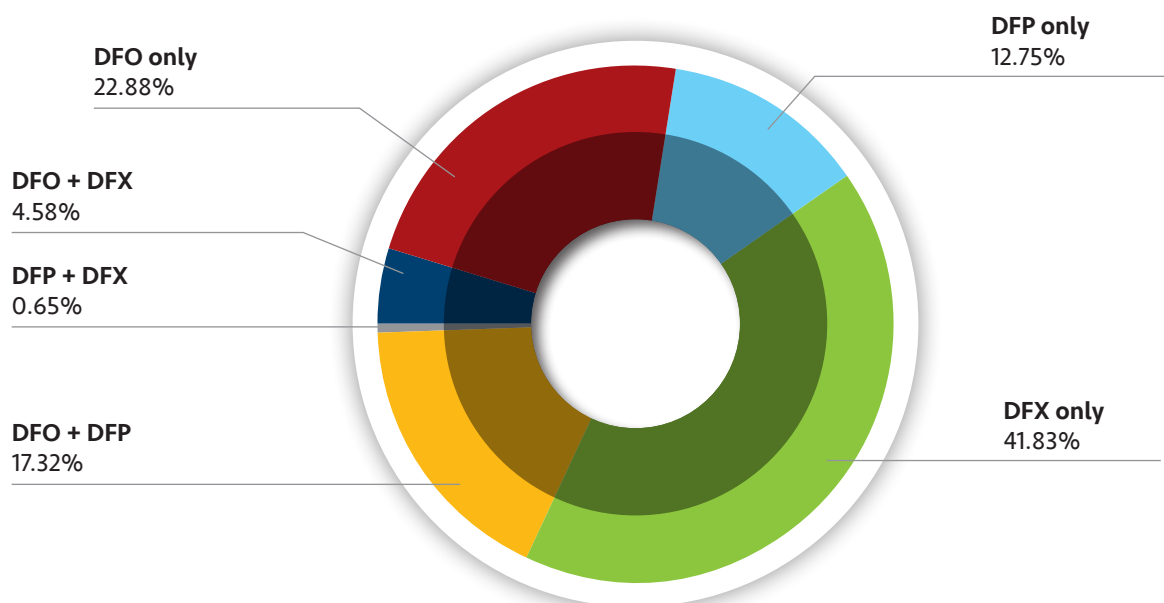


Table 17.14: Distribution of Patients in Kuala Lumpur According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-4.9	14	DFO only	1	0.33
		DFP only	0	0.00
		DFX only	13	4.25
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5-9.9	61	DFO only	9	2.94
		DFP only	1	0.33
		DFX only	46	15.03
		DFO + DFP	1	0.33
		DFP + DFX	0	0.00
		DFO + DFX	4	1.31
		DFO + DFP + DFX	0	0.00
10-14.9	68	DFO only	9	2.94
		DFP only	4	1.31
		DFX only	45	14.71
		DFO + DFP	5	1.63
		DFP + DFX	0	0.00
		DFO + DFX	5	1.63
		DFO + DFP + DFX	0	0.00
15-19.9	65	DFO only	20	6.54
		DFP only	10	3.27
		DFX only	20	6.54
		DFO + DFP	11	3.59
		DFP + DFX	0	0.00
		DFO + DFX	4	1.31
		DFO + DFP + DFX	0	0.00
20-24.9	32	DFO only	13	4.25
		DFP only	5	1.63
		DFX only	1	0.33
		DFO + DFP	11	3.59
		DFP + DFX	1	0.33
		DFO + DFX	1	0.33
		DFO + DFP + DFX	0	0.00

17 Wilayah Persekutuan Kuala Lumpur

292

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
25-29.9	32	DFO only	11	3.59
		DFP only	8	2.61
		DFX only	1	0.33
		DFO + DFP	12	3.92
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30-34.9	22	DFO only	5	1.63
		DFP only	5	1.63
		DFX only	1	0.33
		DFO + DFP	10	3.27
		DFP + DFX	1	0.33
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
35-39.9	9	DFO only	2	0.65
		DFP only	5	1.63
		DFX only	1	0.33
		DFO + DFP	1	0.33
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
40-44.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	0.33
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45-49.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
50-54.9	1	DFO only	0	0.00
		DFP only	1	0.33
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55-59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60-64.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 65	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	0.33
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Total			306	100.00

17 Wilayah Persekutuan Kuala Lumpur

17.4.2 Serum Ferritin Level

There were 256 TDT patients recorded in Kuala Lumpur in 2017 and 2018. There were 64 patients (25.00%) with serum ferritin level lower than 1000 ng/mL in 2017, reduced to 58 patients (22.66%) in 2018. Number of patients with serum ferritin level between 1000-2500 ng/mL is 105 (41.02%) in 2017 and 98 (38.28%) in 2018. On the other hand, number of patients with serum ferritin levels above 2500 ng/mL is 57 (22.27%) in 2017 and 66 (25.78%) in 2018. It can be concluded that number of patients who are compliant with the treatments are slightly decreased in 2018.

Table 17.15: Distribution of Patients in Kuala Lumpur According to Serum Ferritin Level in 2017 by Centre

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
IPHKL	95	36	14.06	40	15.63	14	5.47	3	1.17	2	0.78
PPUKM	69	16	6.25	23	8.98	17	6.64	11	4.30	2	0.78
PPUM	92	12	4.69	42	16.41	26	10.16	11	4.30	1	0.39
Total	256	64	25.00	105	41.02	57	22.27	25	9.77	5	1.95

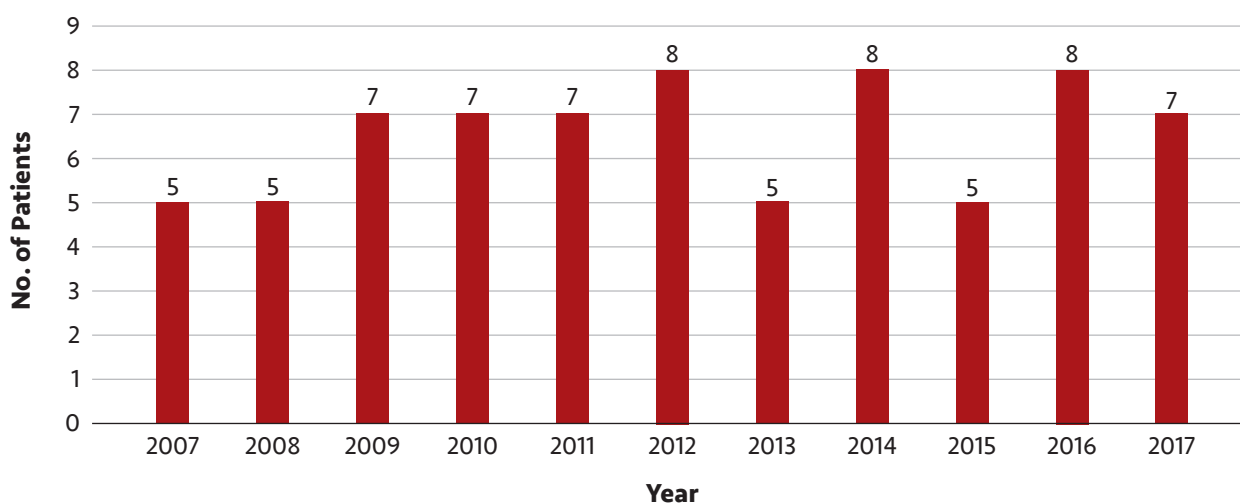
Table 17.16: Distribution of Patients in Kuala Lumpur According to Serum Ferritin Level in 2018 by Centre

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
IPHKL	98	31	12.11	38	14.84	24	9.38	5	1.95	2	0.78
PPUKM	71	18	7.03	20	7.81	16	6.25	14	5.47	3	1.17
PPUM	87	9	3.52	40	15.63	26	10.16	10	3.91	0	0.00
Total	256	58	22.66	98	38.28	66	25.78	29	11.33	5	1.95

17.4.3 Haemopoietic Transplant in Thalassaemia

Since 2007, a total of 77 patients underwent stem cell transplant in IPHKL. The patients were referred from hospitals in all of Malaysia.

Figure 17.7: Number of Thalassaemia Patients Who Underwent Stem Cell Transplant at IPHKL by Year



17.5 Observation and Comment

The report focuses on the result of patient management and outcomes ever since MTR has been operational, from 2007 to 2018. It describes outcomes based on demographic statistics, the types of thalassaemia and effectiveness of treatment.

There are 287 (53.64%) male patients and 248 (46.36%) female patients in Kuala Lumpur. The youngest patient is 2 years old and the oldest is 66 years old. Patients between 10-19.9 years old form the largest number of patients in Kuala Lumpur. Generally, as paediatric patients turn 18 years old, they are transferred to the adult treatment centres (usually Hospital Ampang). Malay patients are the largest group with 393 patients (73.46%), followed by Chinese with 113 patients (21.12%) and Indians with only three patients (0.56%). A total of 11 foreigners are also seeking treatment for thalassaemia in Kuala Lumpur facilities. The treatment fees and iron chelators are self-paid for foreigners, and these cost more than for citizens.

Most children with moderate to severe thalassaemia show signs and symptoms within the first two years of life. If the doctors suspect the child has thalassaemia, he or she may confirm the diagnosis using blood tests. The Hb analysis uses high-performance liquid chromatography (HPLC) test and capillary electrophoresis (CE). HbE-beta thalassaemia and beta thalassaemia major afflicts 237 (44.30%) and 152 (28.41%) of patients in Kuala Lumpur. Another notable diagnosis is HbH disease with 100 patients diagnosed (18.69%), whereas 26 (4.86%) patients have other diagnoses.

Iron chelation therapy is essential for patients with thalassaemia to remove iron overload. In total, 237 patients (77.45%) in Kuala Lumpur are on monotherapy and 69 patients (22.55%) are on combination therapy. The most common combination of chelators are DFO and DFP, prescribed to 53 patients (17.32%). A significant number of patients are on monotherapy with DFX (128 patients, 41.83%).

A total of 169 out of 535 patients (31.59%) have a serum ferritin level lower than 2500 ng/mL in 2017, compared to 156 out of 535 patients (29.16%) in 2018.

18.1 Introduction

Labuan is a federal territory in East Malaysia. It is an island off the shoreline of the territory of Sabah. The Federal Territory of Labuan comprises Labuan Island (75 km²) and six other smaller islands (Pulau Burung, Pulau Daat, Pulau Kuraman, Pulau Papan, Pulau Rusukan Kecil, and Pulau Rusukan Besar), which have a combined total area of 91 km².

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

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

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

18.2 Patient Demographics

Table 18.1: Distribution of Patients in Labuan by Vital Status

Vital Status	Patients
Alive	24
Cumulative Reported Cured by Stem Cell Therapy	1
Lost to Follow-up	0
Total	25
Deaths in 2018	0
Cumulative Reported Deaths	4

Table 18.2: Cumulative Causes of Death Since 2007 in Labuan

Cause of Death	No. of Patients
Health Failure	0
Cardiac Cause	0
Endocrine Complication	0
Infection	1
Other	1
Unknown*	2
Total	4

*Missing data.



18.2.1 Age

From a total of 25 patients in Labuan, 24 are aged between 0-25 years old, as shown Table 18.3.

Table 18.3: Distribution of Patients in Labuan by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0 - 4.9	6	24.00
5 - 9.9	9	36.00
10 - 14.9	5	20.00
15 - 19.9	3	12.00
20 - 24.9	1	4.00
25 - 29.9	0	0.00
30 - 34.9	0	0.00
35 - 39.9	0	0.00
40 - 44.9	0	0.00
45 - 49.9	1	4.00
50 - 54.9	0	0.00
55 - 59.9	0	0.00
60 - 64.9	0	0.00
Above 65	0	0.00
Total	25	100.00

297

MALAYSIAN THALASSAEMIA REGISTRY
REPORT 2018

Figure 18.1: Distribution of Patients in Labuan by Age Group

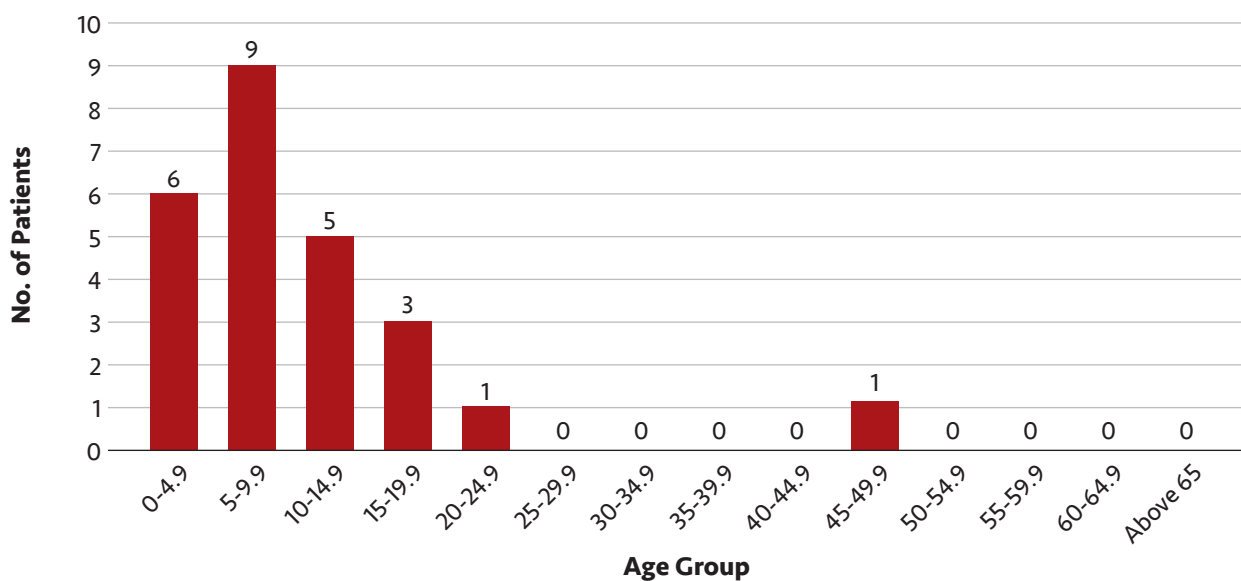


Table 18.4: Distribution of Patients in Labuan According to Age Group by Diagnosis

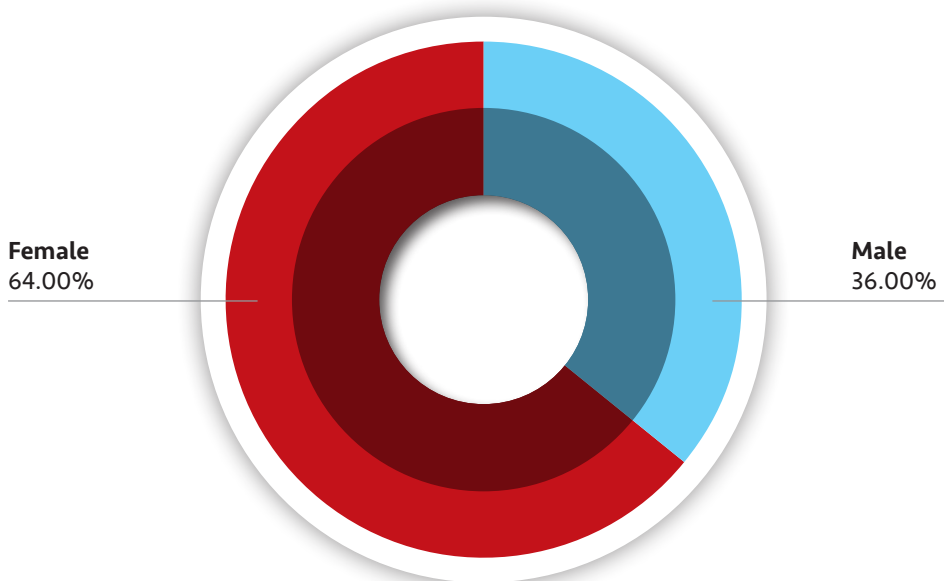
Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0 - 4.9	6	Beta Thalassaemia Major	2	8.00
		Beta Thalassaemia Intermedia	2	8.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	1	4.00
		Others	1	4.00
5 - 9.9	9	Beta Thalassaemia Major	6	24.00
		Beta Thalassaemia Intermedia	1	4.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	2	8.00
10 -14.9	5	Beta Thalassaemia Major	3	12.00
		Beta Thalassaemia Intermedia	2	8.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
15 -19.9	3	Beta Thalassaemia Major	2	8.00
		Beta Thalassaemia Intermedia	1	4.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
20 - 24.9	1	Beta Thalassaemia Major	1	4.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
25 - 29.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
30 - 34.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00

Age Group (years)	Total No. of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
35 - 39.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
40 - 44.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
45 - 49.9	1	Beta Thalassaemia Major	1	4.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
50 - 54.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
55 - 59.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
60 - 64.9	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Above 65	0	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			25	100.00

18.2.2 Gender

Table 18.5 shows the distribution of thalassaemia patients in Labuan by gender. Female patients are higher in number, with 16 patients (64%), and only 9 male patients (36%).

Figure 18.2: Distribution of Patients in Labuan by Gender



300

Table 18.5: Distribution of Patients in Labuan by Gender

Centre	Male		Female	
	No.	%	No.	%
Hospital Labuan	9	36.00	16	64.00
Total	9	36.00	16	64.00

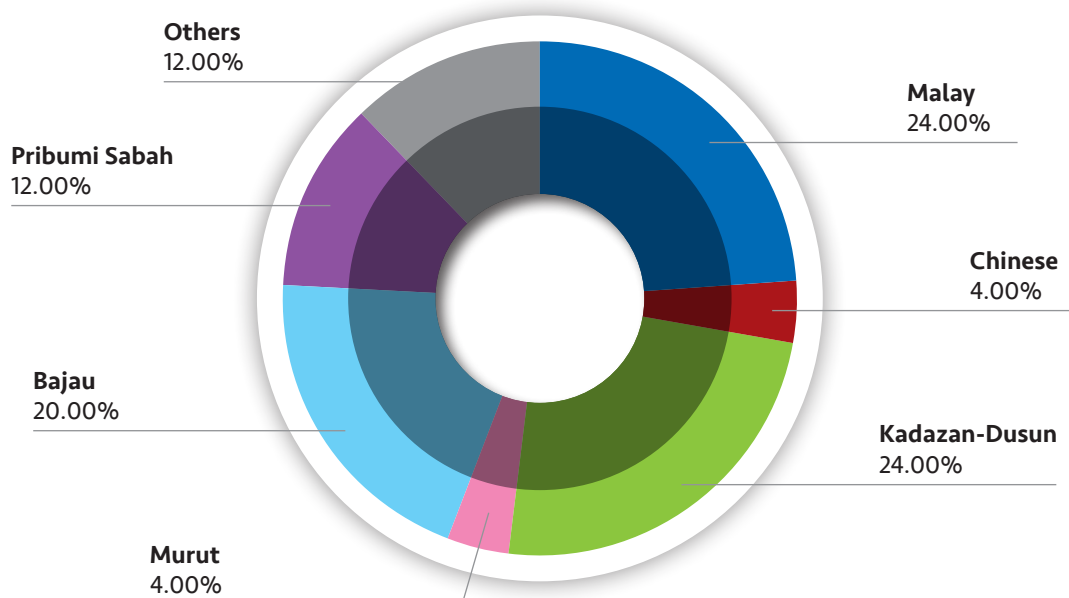
18.2.3 Ethnic Group

The Kadazan-Dusuns and Malays form the largest group of patients in Labuan with six patients (24%) each, followed by Bajau with five patients (20%), Pribumi Sabah and "other" ethnicities with three patients (12%) each, and Chinese and Murut with one patient (4%) each. Each ethnic groups of thalassaemia patients in Labuan is shown in Table 18.6.

Table 18.6: Distribution of Patients in Labuan by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	6	24.00
Chinese	1	4.00
Kadazan-Dusun	6	24.00
Murut	1	4.00
Bajau	5	20.00
Pribumi Sabah	3	12.00
Others	3	12.00
Total	25	100.00

Figure 18.3: Distribution of Patients in Labuan by Ethnic Group



18.3 Diagnosis

Most patients (15 patients, 60%) in Labuan have beta thalassaemia major, followed by beta thalassaemia intermedia with six patients (24%). One patient (4%) was diagnosed with HbH disease, and the remaining three patients (12%) have other diagnoses, as shown in Table 18.7.

Table 18.7: Distribution of Patients in Labuan by Diagnosis

Diagnosis	No. of Patients	Percentage (%)
Beta Thalassaemia Major	15	60.00
Beta Thalassaemia Intermedia	6	24.00
HbE-Beta Thalassaemia	0	0.00
HbH Disease	1	4.00
Others	3	12.00
Total	25	100.00

Figure 18.4: Distribution of Patients in Labuan by Diagnosis

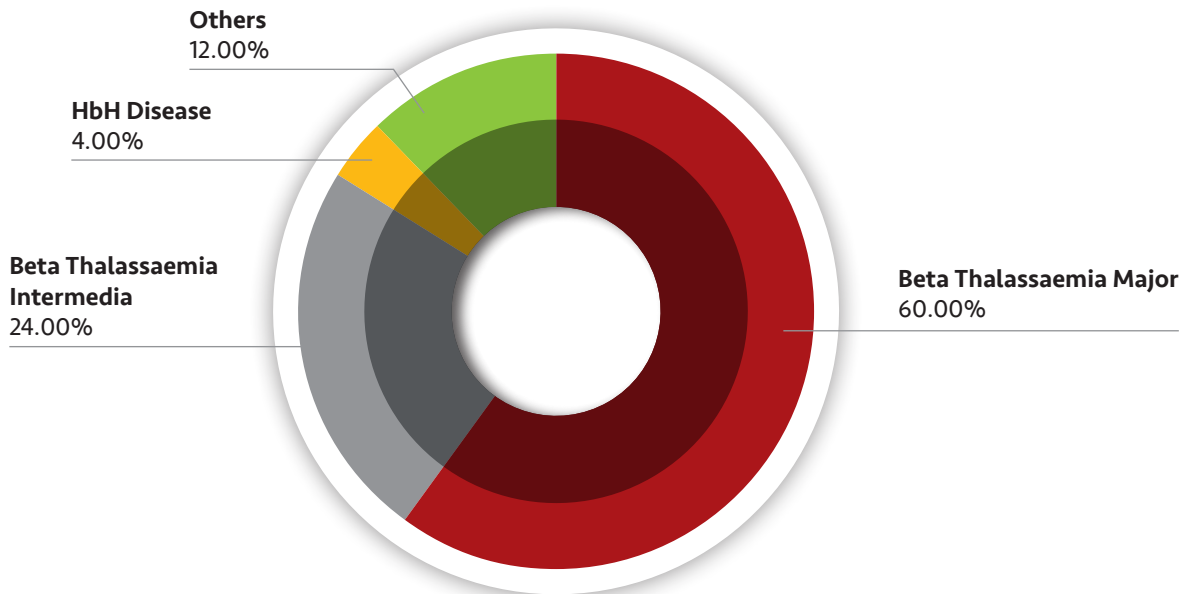


Table 18.8: Distribution of Patients in Labuan According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassemia Major	15	Malay	5	20.00
		Chinese	0	0.00
		Kadazan-Dusun	5	20.00
		Murut	1	4.00
		Bajau	3	12.00
		Pribumi Sabah	0	0.00
		Others	1	4.00
Beta Thalassemia Major	6	Malay	1	4.00
		Chinese	1	4.00
		Kadazan-Dusun	1	4.00
		Murut	0	0.00
		Bajau	1	4.00
		Pribumi Sabah	2	8.00
		Others	0	0.00
HbE-Beta Thalassemia	0	Malay	0	0.00
		Chinese	0	0.00
		Kadazan-Dusun	0	0.00
		Murut	0	0.00
		Bajau	0	0.00
		Pribumi Sabah	0	0.00
		Others	0	0.00

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
HbH Disease	1	Malay	0	0.00
		Chinese	0	0.00
		Kadazan-Dusun	0	0.00
		Murut	0	0.00
		Bajau	1	4.00
		Pribumi Sabah	0	0.00
		Others	0	0.00
Others	3	Malay	0	0.00
		Chinese	0	0.00
		Kadazan-Dusun	0	0.00
		Murut	0	0.00
		Bajau	0	0.00
		Pribumi Sabah	1	4.00
		Others	2	8.00
Total			25	100

18.4 Treatment

18.4.1 Iron Chelation Therapy

Only 18 patients in Labuan are receiving iron chelation therapy. Nine patients (50%) are on DFX, followed by three patients (16.67%) each are on DFO and DFO/DFP combination, two patients (11.11%) on DFP/DFX combination and one patient (5.56%) on DFO/DFP/DFX combination.

Table 18.9: Distribution of Patients in Labuan by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	3	16.67
DFP only	0	0.00
DFX only	9	50.00
DFO + DFP	3	16.67
DFP + DFX	2	11.11
DFO + DFX	0	0.00
DFO + DFP + DFX	1	5.56
Total	18	100.00

Figure 18.5: Distribution of Patients in Labuan by Type of Iron Chelator Received

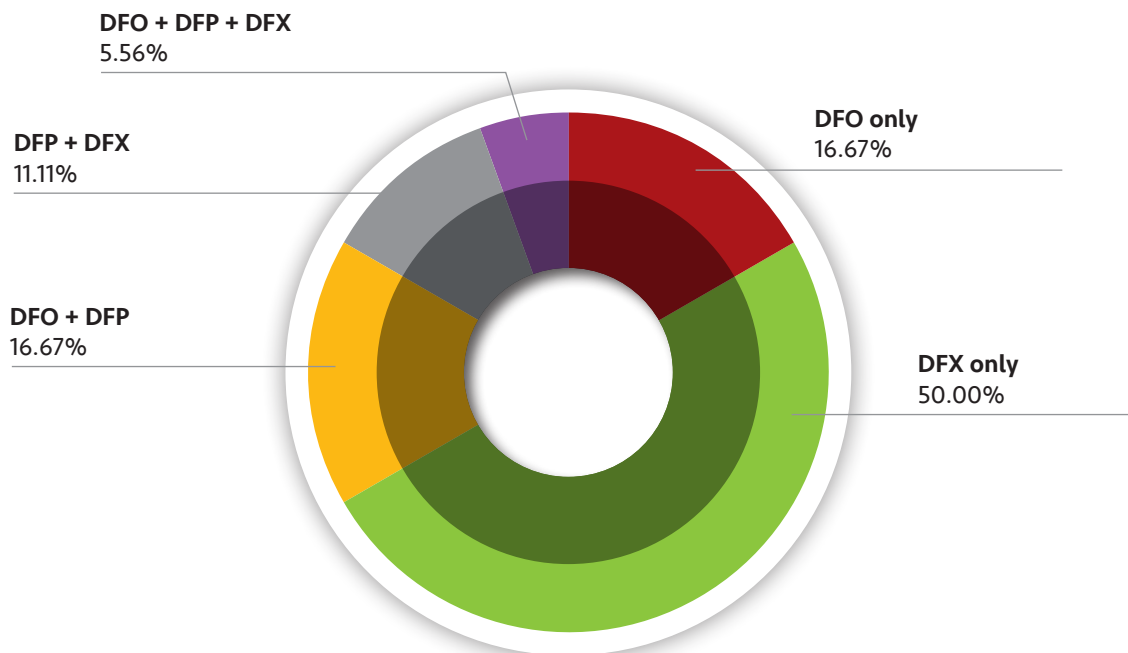


Table 18.10: Distribution of Patients in Labuan According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0 - 4.9	3	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	3	16.67
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
5 - 9.9	6	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	4	22.22
		DFO + DFP	0	0.00
		DFP + DFX	1	5.56
		DFO + DFX	0	0.00
		DFO + DFP + DFX	1	5.56
10 - 14.9	4	DFO only	1	5.56
		DFP only	0	0.00
		DFX only	2	11.11
		DFO + DFP	1	5.56
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
15 - 19.9	3	DFO only	2	11.11
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	1	5.56
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
20 - 24.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	5.56
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
25 - 29.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
30 - 34.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
35 - 39.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

18 Labuan

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
40 - 44.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
45 - 49.9	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	1	5.56
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
50 - 54.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
55 - 59.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
60 - 64.9	0	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	0	0.00
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
Above 65	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			18	100.00

18.4.2 Serum Ferritin Level

Only 12 patients in Labuan have their serum ferritin level measured in 2018. The lowest ferritin level is 195.20 ng/mL and the highest is 13366.00 ng/mL. Two patients (16.67%) have a serum ferritin level less than 1000 ng/mL, three patients (25%) have a serum ferritin between 1000-2499 ng/mL, and six patients (50%) have a serum ferritin level within 2500-4999 ng/mL. Lastly, only one patient (8.33%) has a serum ferritin level higher than 10000 ng/mL.

Table 18.11: Distribution of Patients in Labuan According to Most Recent Serum Ferritin Level

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Labuan	12	2	16.67	3	25.00	6	50.00	0	0.00	1	8.33
Total	12	2	16.67	3	25.00	6	50.00	0	0.00	0	8.33

18.5 Observation and Comments

This report gives a brief overview on the number of thalassaemia patients in Labuan, the types of thalassaemia they have and the basic treatment available for them. Hospital Labuan has only 25 patients with thalassaemia. The most common type of thalassaemia is beta thalassaemia major.

Currently, the patients in Labuan are reviewed by Medical Officer in the thalassaemia clinic with assistance from the visiting Haematologist, Dr Lily Wong from Hospital Queen Elizabeth, Sabah, every three months. Since there is no day care centre in this hospital, all thalassaemia patients receiving treatment will be admitted to ward and follow regular appointment in the clinic. The main problem at this centre is the limited number of experienced nurses in thalassaemia patient management.

18.5.1 Recommendations

1. A standardised chart of thalassaemia and patient diary can be used across the state and be made available for every thalassaemia patient. Thus, data can be easily collected by the RA.
2. To conduct regular healthcare personnel training on thalassaemia management.
3. Staff nurses are recommended to record the date of death and age of deceased patients.
4. Adult patients to have serum ferritin level checked every three or six months.

19 Putrajaya

19.1 Introduction

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

Putrajaya was named after the first Malaysian Prime Minister, Tunku Abdul Rahman Putra. The city is situated within the Multimedia Super Corridor, beside the also newly-developed Cyberjaya. The development began in 1995 and today, major landmarks are completed and the population is expected to grow in the relatively new city. "Jaya" in the city's name means "victory" in Sanskrit.

As of 2017, the population of Putrajaya is around 90,000.

19.2 Patient Demographics

Data analysed were taken from patients with status of either alive, lost to follow-up or cured by transplant, and excludes deceased patients. In total, there are 41 living patients in Putrajaya, as well as one deceased patient.

Table 19.1: Distribution of Patients in Putrajaya by Centre

Centre	No. of Patients (n)	Percentage (%)
Hospital Putrajaya	41	100.00
Total	41	100.00

Table 19.2: Distribution of Patients in Putrajaya by Vital Status

Vital Status	No. Of Patients
Alive	37
Cumulative Reported Cured by Stem Cell Therapy	3
Lost to Follow-up	1
Total	41
Deaths in 2018	0
Cumulative Reported Deaths	1

Table 19.3: Cumulative Cause of Death Since 1997 in Putrajaya

Cause of Death	No. of Patients
Infections	1
Total	1



19.2.1 Age

The youngest patient is 1 year 11 months old and the eldest patient is 44 years old (diagnosed with beta thalassaemia intermedia). The age group with the highest number of patients in Putrajaya is between 10-14.9 years old, with 14 patients.

Table 19.4: Distribution of Patients in Putrajaya by Age Group

Age Group (years)	No. of Patients (n)	Percentage (%)
0-4.9	10	24.39
5-9.9	11	26.83
10-14.9	14	34.15
15-19.9	2	4.88
20-24.9	2	4.88
Above 25	2	4.88
Total	41	100.00

Figure 19.1: Distribution of Patients in Putrajaya by Age Group

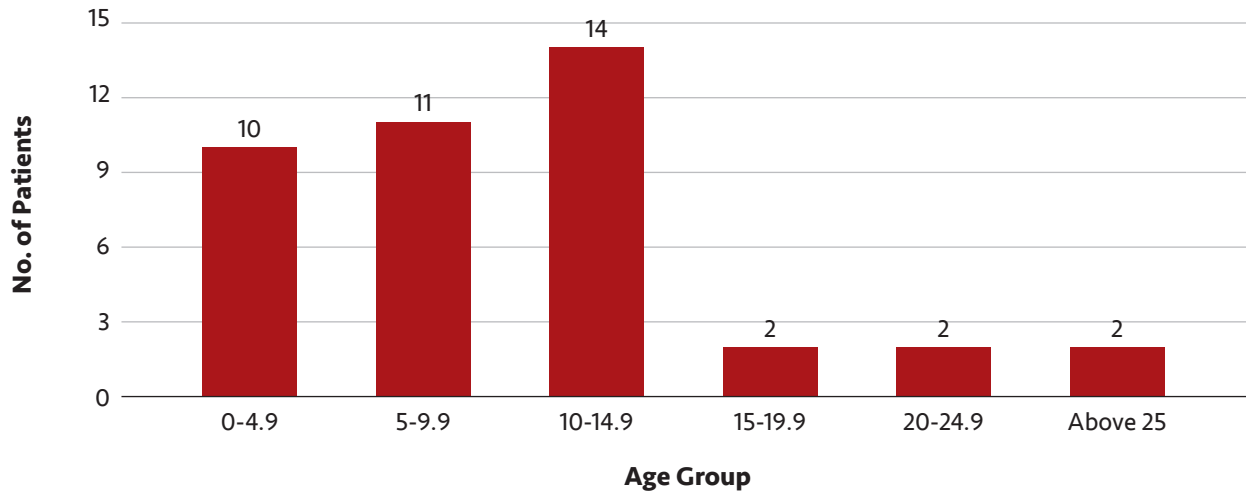


Table 19.5: Distribution of Patients in Putrajaya According to Diagnosis by Age Group

Age Group (years)	Total No. Of Patients	Diagnosis	No. of Patients (n)	Percentage (%)
0-4.9	10	Beta Thalassaemia Major	3	7.32
		Beta Thalassaemia Intermedia	1	2.44
		HbE-Beta Thalassaemia	5	12.20
		HbH Disease	1	2.44
		Others	0	0.00
5-9.9	11	Beta Thalassaemia Major	4	9.76
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	7	17.07
		HbH Disease	0	0.00
		Others	0	0.00
10-14.9	14	Beta Thalassaemia Major	2	4.88
		Beta Thalassaemia Intermedia	3	7.32
		HbE-Beta Thalassaemia	7	17.07
		HbH Disease	2	4.88
		Others	0	0.00
15-19.9	2	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	0	0.00
		HbE-Beta Thalassaemia	2	4.88
		HbH Disease	0	0.00
		Others	0	0.00
20-24.9	2	Beta Thalassaemia Major	1	2.44
		Beta Thalassaemia Intermedia	1	2.44
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Above 25	2	Beta Thalassaemia Major	0	0.00
		Beta Thalassaemia Intermedia	2	4.88
		HbE-Beta Thalassaemia	0	0.00
		HbH Disease	0	0.00
		Others	0	0.00
Total			41	100.00

19.2.2 Gender

Table 19.6 below shows the distribution of patients in Putrajaya by gender. Thirty-two patients (78.05%) are male and 9 patients (21.95%) are female.

Figure 19.2: Distribution of Patients in Putrajaya by Gender

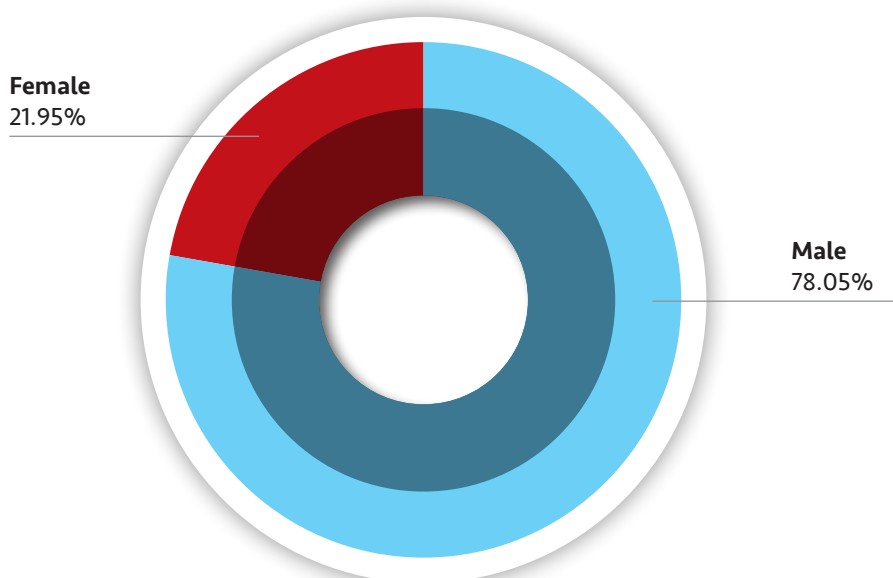


Table 19.6: Distribution of Patients in Putrajaya by Gender

Centre	Male		Female	
	No.	%	No.	%
Hospital Putrajaya	32	78.05	9	21.95
Total	32	78.05	9	21.95

19.2.3 Ethnic Group

Table 19.7 shows that 40 patients (97.56%) in Putrajaya are Malay and one patient (2.44%) is Chinese.

Table 19.7: Distribution of Patients in Putrajaya by Ethnic Group

Ethnic Group	No. of Patients (n)	Percentage (%)
Malay	40	97.56
Chinese	1	2.44
Total	41	100.00

Figure 19.3: Distribution of Patients in Putrajaya by Ethnic Group

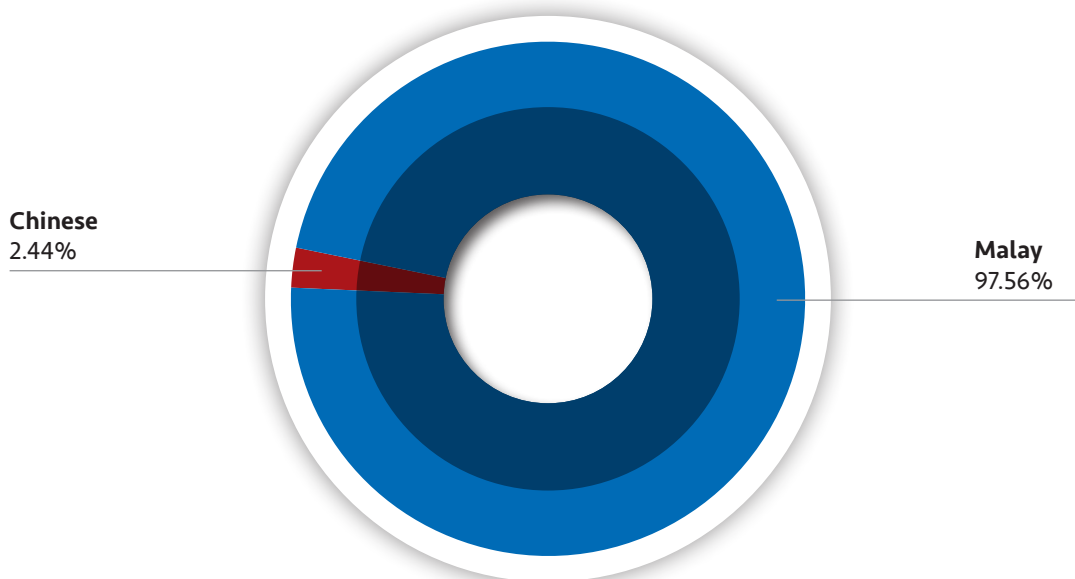


Table 19.8: Distribution of Patients in Putrajaya by Ethnic Group by Centre

Centre	Malay		Chinese	
	No.	%	No.	%
Hospital Putrajaya	40	97.56	1	2.44
Total	40	97.56	1	2.44

19.3 Diagnosis

Half of thalassaemia patients in Putrajaya were diagnosed as HbE-beta thalassaemia (21 patients, 50%), followed by beta thalassaemia major with 10 patients (23.81%).

Table 19.9: Distribution of Patients in Putrajaya by Diagnosis

Diagnosis	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	10	24.39
Beta Thalassaemia Intermedia	7	17.07
HbE-Beta Thalassaemia	21	51.22
HbH Disease	3	7.32
Others	0	0.00
Total	41	100.00

Figure 19.4: Distribution of Patients in Putrajaya by Diagnosis

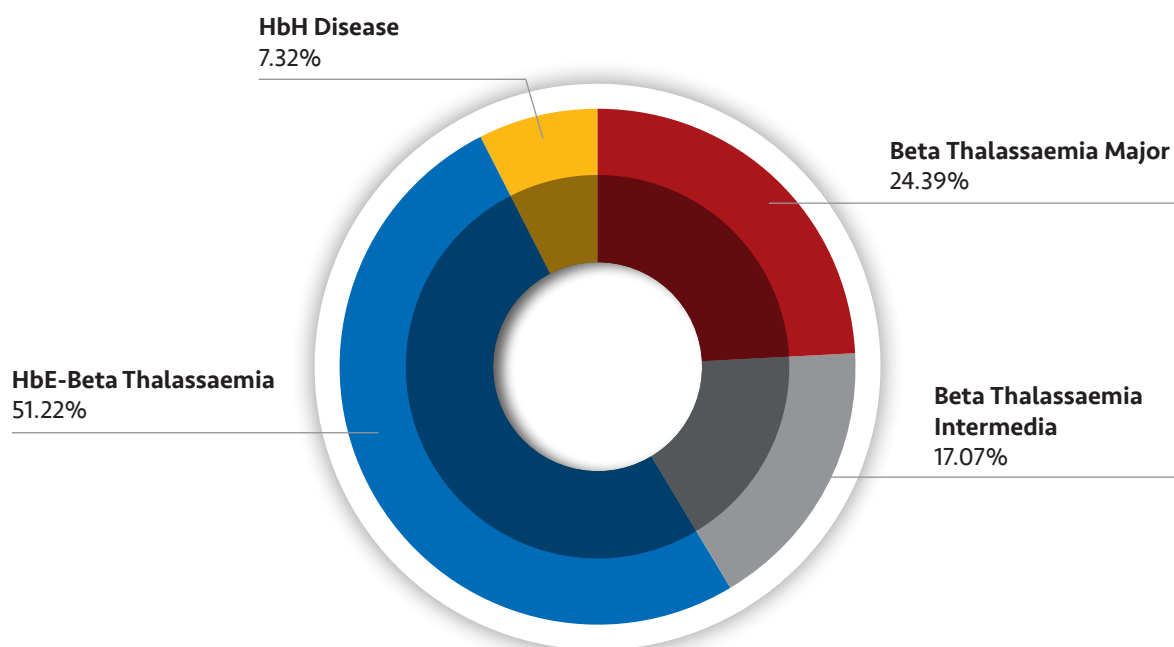


Table 19.10: Distribution of Patients in Putrajaya According to Ethnic Group by Diagnosis

Diagnosis	Total No. of Patients	Ethnic Group	No. of Patients (n)	Percentage (%)
Beta Thalassaemia Major	10	Malay	9	21.95
		Chinese	1	2.44
Beta Thalassaemia Intermedia	7	Malay	7	17.07
HbE-Beta Thalassaemia	21	Malay	21	51.22
HbH Disease	3	Malay	3	7.32
Total			41	100.00

19.4 Treatment

19.4.1 Iron Chelation Therapy

As shown in Table 19.11, 21 patients in Putrajaya receive iron chelation therapy. Fifteen patients (71.43%) are on DFX only, four patients (19.05%) are on DFO and two patients (9.52%) are on DFP. The remaining 20 patients in Putrajaya are not on iron chelation therapy.

Table 19.11: Distribution of Patients in Putrajaya by Type of Iron Chelator Received

Iron Chelator	No. of Patients (n)	Percentage (%)
DFO only	4	19.05
DFP only	2	9.52
DFX only	15	71.43
DFO + DFP	0	0.00
DFP + DFX	0	0.00
DFO + DFX	0	0.00
DFO + DFP + DFX	0	0.00
Total	21	100.00

Figure 19.5: Distribution of Patients in Putrajaya by Type of Iron Chelator Received

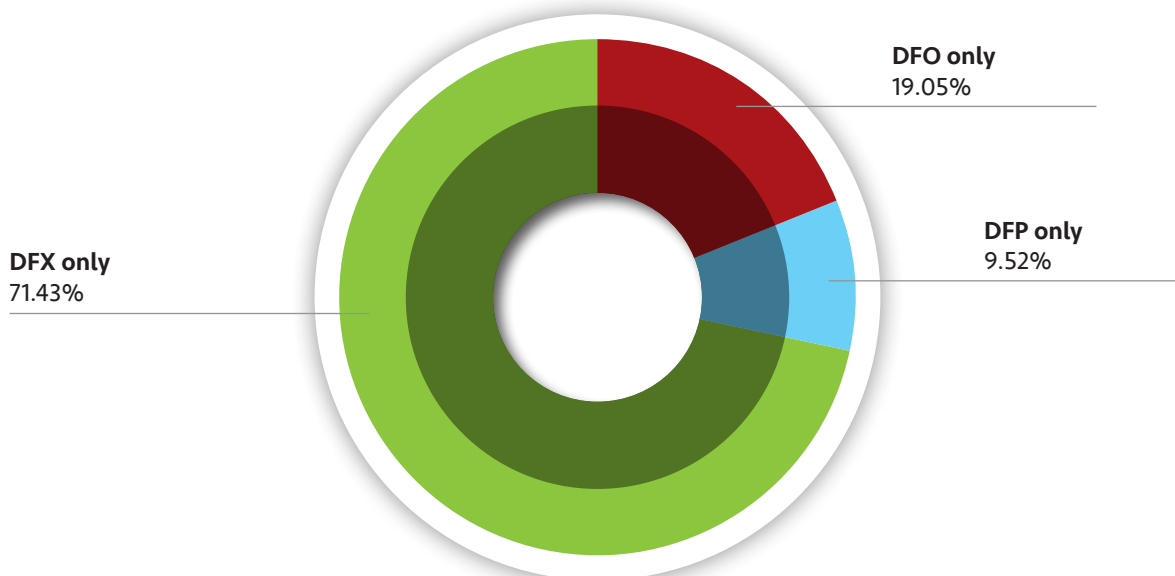


Table 19.12: Distribution of Patients in Putrajaya According to Type of Iron Chelator Received by Age Group

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
0-5	1	DFO only	0	0.00
		DFP only	0	0.00
		DFX only	1	4.76
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
6-10	9	DFO only	2	9.52
		DFP only	0	0.00
		DFX only	7	33.33
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
11-15	7	DFO only	1	4.76
		DFP only	0	0.00
		DFX only	6	28.57
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00

Age Group (years)	Total No. of Patients	Iron Chelator	No. of Patients (n)	Percentage (%)
16-20	1	DFO only	1	4.76
		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
21-25	2	DFO only	0	0.00
		DFP only	1	4.76
		DFX only	1	4.76
		DFO + DFP	0	0.00
		DFP + DFX	0	0.00
		DFO + DFX	0	0.00
		DFO + DFP + DFX	0	0.00
Above 25	1	DFO only	0	0.00
		DFP only	1	4.76
		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			21	100.00

19.4.2 Serum Ferritin Level

The 24 patients on regular transfusion have had their serum ferritin levels measured in 2018. Most of the patients have a serum ferritin level between 1000-2499 ng/mL (20 patients, 83.33%), followed by three patients (12.50%) with serum ferritin level between 2500-4999 ng/mL and one patient (4.17%) with serum ferritin above 5000 ng/mL. These results show that the target serum ferritin level of below 2499 ng/mL have been achieved in more than 80% of patients in Putrajaya.

Table 19.13: Distribution of Patients in Putrajaya According to Most Recent Serum Ferritin Level in 2018

Serum Ferritin Level (ng/mL)		< 1000		1000-2499		2500-4999		5000-9999		10,000+	
Centre	Total	No.	%	No.	%	No.	%	No.	%	No.	%
Hospital Putrajaya	24	6	25.00	14	58.33	3	12.50	1	4.17	0	0.00
Total	24	6	25.00	14	58.33	3	12.50	1	4.17	0	0.00



19 Putrajaya

19.5 Observation and Comments

Out of 41 patients in Putrajaya, 27 of them require regular transfusions. Thirty-two patients (78.05%) are male and 9 patients (21.95%) are female. Most patients in Putrajaya are of Malay descent (40 patients, 97.56%) and only one patient (2.44%) is of Chinese descent. Twenty-one of the patients are children below 10 years of age.

The majority of patients in Putrajaya were diagnosed with HbE-beta thalassaemia (21 patients, 51.22%), followed by beta thalassaemia major (10 patients, 24.39%) and beta thalassaemia intermedia (7 patients, 17.07%). Twenty-one patients (51.22%) receive iron chelation therapy; 15 patients (71.43%) are on DFX monotherapy, followed by four patients (19.05%) on DFO monotherapy and two patients (9.52%) on DFP monotherapy.

Based on serum ferritin level record in 2018, majority of patients (20 out of 24 TDT patients, 83.33%) in Putrajaya have a serum ferritin level below 2500 ng/mL. The remaining four patients had serum ferritin levels above 2500 ng/mL. Data on serum ferritin levels in Putrajaya were entered only for TDT patients in the registry.

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