Original Article

Frequency of Admission with Hereditary Blood Diseases in Paediatric Wards of Basrah Hospitals Iraq

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ABSTRACT

Objective: To Identify the Causes & Patient Characteristics of Hereditary Blood Diseases of Basrah City Iraq.
Study Design: Retrospective study
Place and Duration of Study: This study was conducted at the Basrah Medical College, Iraq from 1st July 2022 to 31st December 2023.
Methods: This retrospective study reviewed patient records from the hematology ward of a hospital and found that 1,799 inherited hemoglobin disorder patients.
Results: All patients were under 15 years of age, with the majority (34.5%) in the 6–10 years. Of the total patients, 55.48% were male, resulting in a male-to-female ratio of 1.24. Sickle cell–related complications and morbidity were the predominant causes of admission, accounting for 64.48% of the cases. Outpatient referrals constituted the primary pathway for admission, with approximately half of the cases originating from the center and 97.44%. Most patients (88.6%) had a hospital stay of fewer than five days. Only 0.67% (12 patients) had prolonged stays (exceeding 15 days), primarily due to sickle cell–related morbidities. The average length of stay was 2.003 days, the range being 0–35 days.
Conclusion: Most of the admission trend was toward sickle cell and complications and recommendations were toward more preventive efforts toward vasoclosive pain and other complications in sickle cell patients
Key Words: Hemoglobinopathies, Sickle cell disease, Thalassemia, Inpatient, Admission

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INTRODUCTION

Sickle cell disease and thalassemia stand as the principal well-known inherited blood abnormalities which affect broad regions of Mediterranean and Asian and African lands because of endemic malaria conditions.[1] Sigma 6 HBB point mutation causes sickle cell disease by creating beta-globin chain amino acid substitution at position 6. These mutations occur on autosomal recessive chromosomes. Sickle cell disease stands as one of the most common hemoglobin disorders found in Saudi Arabia and especially across its population.[2] Sickle cell disease happens with five unique beta-globin gene haplotypes which include the patterns from Central African Republic, Cameroon, Benin, Senegal and Arab-Indian. HbF production is directly influenced by genetic factors which work to manage sickle cell disease severity because higher levels of HbF usually result in milder disease symptoms.[3,4]

The inherited hemoglobin disorders known as β-thalassemia causes significant health care challenges to Europe and worldwide because of their complicated nature and diverse characteristics. Genetic studies demonstrate that β-thalassaemic mutations exist in 1% to 5% of people worldwide.[5,6] Therapeutic and medical advancements significantly impacted the survival rates of patients with SCD particularly among young children because they increased the chances of surviving the diseases and outliving the original death prediction. A large majority (93.9%) of sickle cell anemia children and 98.4% of children with less severe SCD disease forms achieve adult life.[7] In Basrah, the prevalence of sickle Hemoglobin (Hb S) is 6.48%, with a carrier frequency of 0.0324%.[8]

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children with less severe SCD disease forms achieve adult life.[9] Therapeutic and medical advancements significantly impacted the survival rates of patients with SCD particularly among young children because they increased the chances of surviving the diseases and outliving the original death prediction. A large majority (93.9%) of sickle cell anemia children and 98.4% of children with less severe SCD disease forms achieve adult life.[10] The total number of people who had hemoglobinopathies amounted to 238 per 100,000 while the male population had 247 per 100,000 and the female population had 229 per 100,000. Most patients in the study belonged to the age group of 6 to 15 years and were male at 52.11% of the total sample. The number of sickle cell disease patients accounted for 68.58% of total cases. Sickle crisis appeared most frequently among reported complications which amounted to 14.21% of total cases while sickle crisis itself occurred in 26.83% of patients. Since 2012 the number of cases reached its maximum at 606 during 2019 then subsequently reduced to 349 in 2022.[12] The purpose was to determining how often children with various hemoglobinopathies and associated haematological disorders need hospital admission care along with identifying their respective causes in Basrah. Findings from this research will build a necessary understanding of thalassemia health problems in the country which will help create preventive measures for managing severe complications in affected individuals.

METHODS

This observational study that depend on review of the patient records to collect certain demographic and clinical data, Demographic data for each patient include age (below 5 years, 5-10, 11-15), sex, locality (within Iraq, within Basrah), main disease category (sickle related, thalassemia related, bleeding disorders related and other haematological categories), and referral point (outpatient clinic, day care clinic, other hospital or emergency room). Clinical data include duration of admission, cause of admission and patient outcome (improved, discharge on responsibility, died or transferred to other hospital or centre). All the admitted haematological case inpatient of the BCHBD (the point at which the centre transferred to the current location within the Basrah paediatric speciality teaching hospital). The data was entered and analyzed through SPSS-22. P value by linear ANOVA had been calculated.

RESULTS

Mostly of the patients 682 were 6-10 years, 55.48% were males with a male to female ratio of 1.24, sickle cell related complications and morbidity were the most predominant 64.48%, outpatient referral were the most path for admission, and about half of the cases were from the centre while 97.44% were from Basrah governorate, 45.9% of the whole cohort study. Minimum 22 patients in August 2022, maximum 201 patients are in November 2023 with an average of 105 during the study period (Table 1).

The average duration of admission was 2.003 days. Most of the admitted cases were of a duration <5 days 1594 (88.60%) while only 12 (0.67%) were of prolonged admission stay (>15 days) in which most of them were of sickle cell related morbidities and average of the admission duration was 2.003 day (Table 2)

Table No.1: Demographic characteristics of the admitted patients

Variable	No.	%
Age (years)		
1-5	541	30.4
6-10	682	37.9
10-15	569	31.7
Gender		
Male	998	55.8
Female	797	44.2
Main categories		
sickle cell related	1160	64.4
Thalassemia related	258	14.5
bleeding disorder related	212	11.7
Others	169	9.4
Referral		
Outpatient	1017	56.5
Day clinic	543	30.3
Other hospital	34	1.8
Privates	205	11.4
Locality		
Basrah	1753	97.4
Other governorates	46	2.6
Locality within Basrah		
Centre	896	49.8
Abulkhasseb	238	13.2
Zubair	148	8.2
Fao	18	2.0
Safwan	7	0.4
Qurna	71	4.9
Mudynaah	65	4.6
Hartha	79	4.4
Shat Alarab	231	12.5
Time trend of admission		
2022		
July-September	46	2.6
November-December	263	14.6
2023		
Jan-March	406	22.6
April-June	420	23.4
July-September	416	23.3
November-December	247	13.7

The most common isolated cause for in patient admission in Basrah was sickle cell vaso-oclosive crises (40.19%) followed by transfusion emergencies and

bleeding in a patient with clotting disorders (20.68, 10.78%) respectively (Table 3). Patients being improved, discharged on responsibility, referred to other hospital or died the patient distribution were 62.37%, 36.24%, 0.50% and 0.44% respectively (Table 4).

Table No.2: Duration of admission categories

Duration of admission			Sickle cell related		Thalassemia related		Bleeding disorder related		Others	
0-4 days	1594	88.60%	1022	88.10%	226	87.60%	201	94.81%	145	85.80%
5 - 9days	162	9.01%	107	9.22%	23	8.91%	10	4.72%	22	13.02%
10-14 days	31	1.72%	23	1.98%	6	2.33%	1	0.47%	1	0.59%
15 & above	12	0.67%	8	0.69%	3	1.16%	0	0.00%	1	0.59%
Total	1799	100%	1160	64.48%	258	14.34%	212	11.78%	169	9.39%

***0 mean less 24 hours**

Table 3: Causes of admission categories

Causes of admission		No.	%
Sickle related	Sequestration crises	114	6.34
	Acute chest syndrome	25	1.39
	Pulmonary infection	48	2.67
	Sickle painful crises	723	40.19
	Febrile sickle illness	61	3.39
	UTI	19	1.06
	Musculoskeletal infection	23	1.28
	Aplastic crises	3	0.17
	Stroke	5	0.28
	Other infectious diseases	69	3.84
	Sickle nephropathy	3	0.17
Thalassemia related	Blood transfusion emergency	372	20.68
	Hormonal disturbance and complication	9	0.50
Bone marrow failure related		42	2.33
Bleeding disorders related	Clotting	194	10.78
	Platelets	37	2.06
HLH		13	0.72
Preparation to surgery		8	0.44
Other causes		31	1.72

Table No. 4: Fate of the admitted cases

Fate of the patients	No.	%
Improved	1122	62.37
Discharged on responsibility	652	36.24
Referred to other hospital	9	0.50
Death	8	0.44

DISCUSSION

The sex distribution and sickle cell reasons for hospital stay were found to be comparable between this study and another Saudi research design. Researchers studied 103 SCD patients through an age range of 18–62 years. The majority of sickle cell disease patients at the facility were males who belonged to the ≤30 year old age group. Most admissions for sickle cell patients were associated with a painful crises (n=94, 91.3%) followed by cases of hemolytic crisis (27 of the cases; 26.2%) and acute chest syndrome (n=32, 31.1%) exams.[13]

The Lebanese series investigation demonstrated thalassemia patients needed transfusions in fifty-four percent of cases while fewer patients received treatment for infectious conditions. The hospital stay duration exceeded five days within this cohort. The different age range of patients between studies partly explains this finding since elderly adults made up most of the Lebanese data.[14]

The research conducted by Karemi et al during 2011 in Iran demonstrated that males accounted for (59.75%) of the total patient population while females made up (40.25%). Among admitted patients the most common reasons for admission involved splenectomy (21.8%) and infections aetiology (19.9%) and diabetes mellitus (13.4%) and congestive heart failure (19.0%) and Liver biopsy (11.5%); those patients had an average age of 11.28 years.[15]

Sickle cell related morbidity and complications served as the most common hospitalization diagnosis (64.48%) according to this study while a large USA dataset showed 19,250 sickle cell patients had sickle cell-related admissions throughout 2016 to 2020. The annual rate of vasoclosive cause of pain admissions rose from 10.2 per 100,000 individuals in 2016 to 14.7

in 2019 followed by a decrease to 13.9 in 2020. The complications of acute kidney injury presented were 4.9% of admissions. During 2017 the number of bacteraemia illnesses rose to 0.9% while priapism maintained steady growth of 2.3% in 2016 but admissions complicated by pneumonia reached 6% in 2016. Less than one percent of total admissions involved complications of osteomyelitis, heart failure exacerbation, stroke, splenic sequestration, acute chest syndrome, or sepsis and statistical analysis did not show any relevant findings. This cohort demonstrates a female predominance over males The opposite results stem primarily from two factors: the different genotypic categories of SCD and population differences between patients.[16]

A study in Thailand showed high hospital admission rate among paediatric patients with thalassemia under the NHC scheme in Thailand from 2015 to 2019. The admission rate, ranging from 416 to 559 per 100,000, within this cohort, main age category was 11-16 years and minimal was in below 1 year. Most hospitalized patients had β-thalassemia, with iron overload and infections as prominent co-diagnoses. This study identified a significant presence of cardiovascular complications and diabetes mellitus among patients, Respiratory tract infections constituted approximately 63% of all infections.[17] Beazrkar et al[18] in Iran described the causes of hospital admission in 555 patients with beta-thalassemia major at a referral university hospital in Iran from 2000 to 2005. The most common causes of hospital admission were splenectomy, heart failure, liver biopsy, uncontrolled diabetes mellitus, and arrhythmia. Origa et al[19] in Sardinia described 690 hospital admissions in 276 paediatric and adult patients with thalassemia major in a tertiary care centre. The most common causes for hospital admission were heart failure/arrhythmias, infections, mesenteric lymphadenitis in patients treated with subcutaneous desferrioxamine, digestive tract diseases, and liver diseases.

Outpatient referral were the most path for admission, and about half of the cases were from the centre while 97.44% were from Basrah governorate, 45.9 % of the whole cohort study with a monthly admission. A similar trend found in a study in USA in which Hospital admission rate rose steadily from 106 per 100,000 AA populations in 2004 to 137 in 2012. Seasonal and trend decomposition revealed the highest hospitalization rate in January. Hospital LOS decreased from 7.1, 7.65 days in 2004 to 6.23 6.42 days in 2012.[20] While in a large study in UK showed also a 74.9% of admitted were from the capital and 57.9% of patients admitted are discharged within 24 hours referral from ER (79.6) and being from the centre (92.8) area (metro covered areas) was found in a study done by Kathryn et al also the vaso-oclosive pain (78.3%) in this study was the predominant cause in sickle cell patients.

Whether being improved, discharged on responsibility, referred to other hospital or died the patient distribution was 62.37%, 36.24%, 0.50%, 0.44% respectively a similar results were seen in other studies like in US series.[21,22]

CONCLUSION

Most of paediatric admitted patients were from age group 6-10, males referred from outpatients live in the centre of the Governorate. The most disease category was the sickle related type and the most common cause for admission was the vaso-occlusive crises. Average of the admission duration was 2.003 day and most of the long admission duration was sickle cell category. Death was a minority while cure was the most of the admitted patient fate.

Recommendations

1. More interest should concentrate on the sickle cell preventive program to minimize admission rate like vaccinations, antibiotic prophylaxis and regular outpatient visits.
2. More interest should be directed toward the prevention of vaso-occlusive crises like offering hydroxycarbemide and erythrocytapheresis.
3. A more analytic study to be done on the same sample in regards to the risk factor for in patient admission and death fate, and ICU referral

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