To Find Out the Prevalence of Thalassaemia in Sukkur Pakistan

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Abstract

Pakistan is populated to approximately 225,633,392 people (225 million). Pakistan's healthcare delivery system is complex due to the presence of both federal and provincial healthcare subsystems. Iron deficiency anemia and co-pathological diseases like - or -thalassemia (Alpha thalassemia, Beta thalassemia characteristics) are the most frequent disorders among microcytic hypochromic anemias.

More than 10 million people in Pakistan are carriers of the beta-thalassemia trait; every year, around 5,000 children in Pakistan are identified as being carriers of beta-thalassemia major. Despite the widespread use of blood transfusions, there are no established guidelines for patient care. Families tend to be big, and the majority of the population lives in poverty, thus they cannot afford the high costs associated with caring for a kid with thalassemic disorders. This prospective observational research of thalassemia was carried out in a blood bank in the city of Sukkur in the province of Sindh, Pakistan. The purpose of this research was to determine how common thalassemia is in the sukkur area of Sindh, Pakistan. Blood Bank Sukkur was the site of the whole research, which was led by Sajida Mahmood Gopang and included the efforts of Mazhar Hussain kharal, farhan Ali, Ahsan, Sajjad Hussain, and Zain-ul-Abdin. The research concluded that in order to eradicate thalassemia in Pakistan, health care practitioners should be incentivized to raise awareness of the disease among the general population and treat it as a public health issue.

Keywords: *Pakistan; prevalence; thalassemia prevention; hemoglobin (Hb) disorders; β-Thalassemia.*

Introduction

Patients who have thalassemia major have anemia a result of this genetic hematological condition. Chromosome 11 is implicated in this autosomal recessive illness. It has an impact on hemoglobin production, which is either reduced or missing, causing early red blood cell turnover (RBC) (Hajimoradi, 2021). Thalassemia minor occurs when one of the beta globin chain genes is normal and the other is defective. Thalassemia intermedia is the name used when both genes are active and the condition manifests late in life (in older children and adults). Thalassemia major, a more severe condition, is the name given when both genes are implicated and the ailment manifests in a young child or newborn (Hromić-Jahjefendić, 2022). The reticuloendothelial system quickly destroys abnormally shaped RBCs, especially in the spleen, which causes microcrystal hypochromic anemia and iron excess (Isa,2019). Thalassemia patients have anemia a result of their bodies not producing enough hemoglobin. Thalassemia patients cannot make red blood cells on their own and must often have blood transfusions in order to survive. It is an illness that is innate rather than contagious and is unaffected by breathing, eating, sleeping, or living, yet it is passed from parents to children (Mishra, 2022). The most prevalent genetic condition, thalassemia major, poses a serious genetic challenge to people across the globe (Amjad, 2020).

Thalassemia is a rare but serious health issue in Pakistan, where an estimated 5–6 million children have the disease or its symptoms (Ullah, 2022), where over 5,000 Thalassemia homozygotes are born every year 25 (Batool, 2022), and where 6% of the population carries the genes for the disorder because of abnormal production of hemoglobin (Hassan, 2020). Saif-ur-Rehman, (2002) reported that most parents with Thalassemia traits never come to know that they are carrying the genes as Thalassemic carriers, which are the main source of spreading the disease, so these numbers are alarmingly rising due to lack of awareness and insufficient educational campaigns. Because of stigma and concerns about how it would affect their romantic possibilities, many individuals avoid being examined for diseases that can only be detected by blood examination. (Septyana, 2019; CHUSRI, 2019). In addition to the symptoms already associated with sickle cell anemia, individuals also experience a host of additional health issues due to the condition, including fatigue, pale skin, and Thalassemic

features (Chaloemwong, 2019; Kaewkong, 2020). Helplessness, rejection, loss, and sadness are among emotions that may be triggered by the disease's impact on the brain (Maes and Stoyanov, 2022, Maes, 2022). Certain populations, including Punjabis, Sindhis, Gujaratis, Bengalis, Parsis, etc., have a disproportionately high prevalence of thalassemia.

The frequency is about 15% among Punjabis who have emigrated from West Pakistan. The incidence of thalassemia is highest in the northern and eastern regions of India, and lowest in the southern region. A higher prevalence of hemoglobin E illness and hemoglobin E disease in conjunction with thalassemia is seen in the Northeast. As cited in (Pyae, 2019).

The mean age of the population was 15.06 years with a range of 328 years, according to a study conducted by (Chen, 2020) on the Sindhi people who live in and around the city of Wardha in India. Those aged 12-18 made up the bulk of the population. Nestroft reports a 36.36 percent incidence and a prevalence of 200/550 for the carrier condition of thalassemia.

There was a viral (Hadipour Dehshal, 2019) According to HbA2 measurement, 17.2 percent (95/550) of the Sindhi population in the Jamnagar area of Gujarat, India, reported having hemoglobinopathies. The beta thalassemia trait (BTT) was more prevalent in males and females between the ages of 12 and 21, and samples were taken from 109 healthy participants in the Jamnagar district of Gujrat between January and February 2015. There were 21 people out of every 100 who tested positive for hemoglobinopathy (19.26 percent), with 17 of them having beta thalassemia trait (15.59 percent).

(Ul Hassan and Manzoor, 2020). To determine the prevalence of hemoglobinopathies in Hyderabad, Sindh, researchers discovered that the mean age of patients was 8.11 + 03.36 years. Thalassemia major and minor were extremely prevalent, occurring in 36.5 percent (n=301) and 47.5 percent (n=301) of the population, respectively. Sickle cell disease and trait, Hb-D, Hb-C, Hb-D disease, and B-thalassemia with compound heterozygous were next, with percentages of 1.3 percent, 7.6 percent, 0.7 percent, 1.7 percent, 0.7 percent, and 3.3 percent, respectively. Hb-E disorder was also observed.

(Shah, 2020) According to reports, one of the most prevalent hereditary hemoglobin diseases in Pakistan is -thalassemia. 5.4 percent or so of the frequency is the carrier. They examined a representative sample of 602 alleles from six ethnic groups in Pakistan to discover the range of -globin gene abnormalities that cause -thalassemia; 99.2% of the alleles were defined, while 0.8 percent remained unexplained. There are 19 distinct mutations over the whole range of mutations, which is heterogeneous. Eighty-seven percent of the alleles are affected by the four most frequent mutations, IVS-I-5 (GC) (37.7%), codons 8/9 (+G) (21.1%), the 619 bp deletion (12.4%), and IVS-I-1 (GT) (9.5%). Both across provinces and amongst ethnic groupings, there exist disparities. In Pakistan's four provinces, Sindh and Baluchistan, which border India in the south and Iran in the southwest, respectively, have higher rates of the IVS-I-5 (GC) mutation than do Punjab and the Northwest Frontier Province, which border India in the northeast and Afghanistan, respectively, with higher rates of the codons 8/9 (+G) mutation. Gujaratis and Memons living in the Province of Sindh, which is next to the Indian Gujrat, have a greater (619 bp deletion) rate (46%) than other ethnic groups.

Material and methods

This prospective observational research of thalassemia was carried out in a blood bank in the city of Sukkur in the province of Sindh, Pakistan. The purpose of this research was to determine how common thalassemia is in the sukkur area of Sindh, Pakistan. Blood Bank Sukkur was the site of the whole research, which was led by Sajida Mahmood Gopang and included the efforts of Mazhar Hussain kharal, farhan Ali, Ahsan, Sajjad Hussain, and Zain-ul-Abdin. Blood transfusion information was gathered from the parents of thalassemia patients whose information was on file at the Sukkur Blood Bank. data were collected only from patients' parents. A total of 593 questionnaires were used to gather the data; questions pertaining to blood type, illness, gender, symptoms, and family history were the most often asked. Percentages, frequency distributions, means, and standard deviations were calculated using SPSS (Statistical Package for the Social Sciences) version 20 to examine the data.

Results

Frequency tests 1

Under frequency test 1, we conduct the test using data on the total number of patients, their ages, genders, blood types, causes, symptoms, and consequences.

At the blood bank in Sukkur, the research was carried out between the months of August 2022 and November 2022. 320 patients ranged in age from 1 to 10 years old. 200 patients were between the ages of 11 and 20. And 73 of the patients were between the ages of 21 and 25. Most of the 320 patients in the age range of 1 to 10 had a high valid percentage of 53.96 percent. (Table 1)

Ages	*Number*	*Percentage*	*Valid percentage*	*Cumulative percentage*
1 –10	320	53.96%	53.96%	100%
11--20	200	33.727%	33.727%	100%
21--25	73	12.310%	12.310%	100%
TOTAL	593	100%	100%	100%

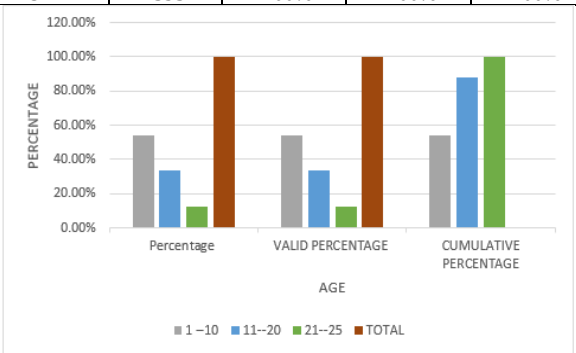


Table 2 displays the gender distribution of thalassemia patients. The total number of males suffering is 385 out of 593 with a valid percentage of 64.925 percent, while females rank second with a valid percentage of 35.075 percent. The cumulative proportion is 100 percent overall.

Genders	*Numbers*	*Percent*	*Valid percentage*	*Cumulative percentage*
Males	385	64.924%	64.924%	100%
Females	208	35.075%	35.075%	100%
TOTAL	593	100%	100%	100%

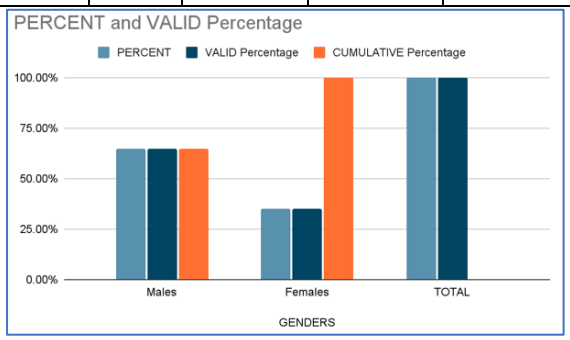


Table 3 shows the distribution of thalassemia patients by blood type. According to the findings of blood bank sukur in 2022, those with the A+ blood group suffer the most, with

a count of 220 and a valid percentage of 37.099%. People with the blood type O+ who have thalassemia rank second with a count of 100 and a valid percentage of 16.864%. 100 percent is the total cumulative percentage of this frequency result.

Blood groups	*Number*	*Percentage*	*Valid percentage*	*Cumulative percentage*
A+ve	220	37.099%	37.099%	100%
A-ve	25	4.216%	4.216%	100%
B+ve	80	13.490%	13.490%	100%
B-ve	40	6.746%	6.746%	100%
AB+ve	60	10.119%	10.119%	100%
AB-ve	23	3.879%	3.879%	100%
O+ve	100	16.864%	16.864%	100%
O-ve	45	7.589%	7.589%	100%
TOTAL	593	100%	100%	100%

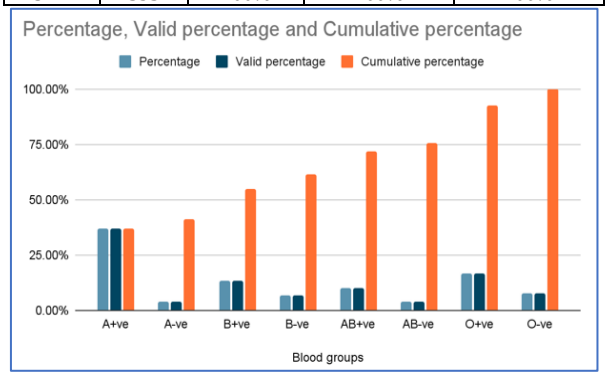


Table 4 displays the prevalence of thalassemia causes in prospective marriages. The data indicate that the majority of patients are the product of cousin marriages, with 553 patients and a valid rate of 93.255 percent. In contrast to cousin weddings, which have a legitimate proportion of 6.75 percent, out-of-family marriages have a low ratio. The cumulative total percentage is 100 percent.

Causes	*Number*	*Percentage*	*Valid percentage*	*Cumulative percentage*
Cousins	553	93.255%	93.255%	100%
Out of family	40	6.745%	6.745%	100%
Total	593	100%	100%	100%

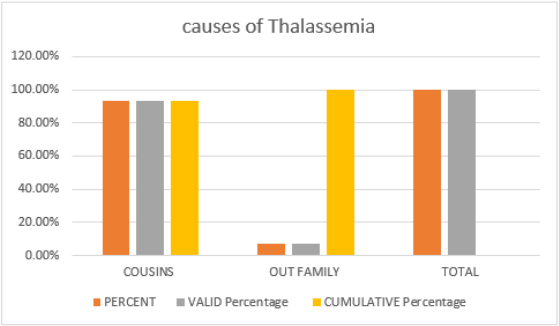


Table 5 displays the findings of the Symptoms of thalassemia patients survey. According to the data, 355 out of 593 individuals with abdominal edema had a valid percentage of 59.865 percent, or a high no. Patients with weakness as a symptom number 178 and have a valid percentage of 30.017%, according to the data; this is the second highest number. Overall, the cumulative proportion is 100 percent.

Symptoms	*Number*	*Percentage*	*Valid percentage*	*Cumulative percentage*
Yellowish Skin	60	10.118%	10.118%	100%
Weakness	178	30.017%	30.017%	100%
Abdomen Swelling	355	59.865%	59.865%	100%
TOTAL	593	100%	100%	100

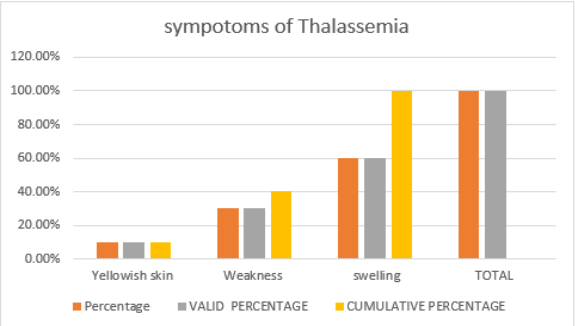
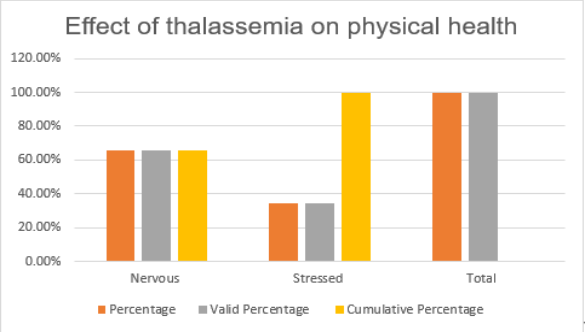


Table 6 displays the physical health implications of thalassemia. With a high valid percentage of 65.430 percent, 388 patients out of 593 believe that anxiousness is the principal impact on them. 205 patients believe that

stress is the other consequence they experience. This result's cumulative percentage is 100 percent.

Physically	*Number*	*Percentage*	*Valid Percentage*	*Cumulative Percentage*
Nervous	388	65.430%	65.430%	100%
Stressed	205	34.569%	34.569%	100%
Total	593	100%	100%	100%



Frequency Test 2

In frequency 2 testing, the ratio of total deaths among thalassemia patients is analyzed. For this aim, we conduct the test with 265 questionnaires.

Table 7 displays the age distribution of thalassemia deaths. With a valid percentage of 71.698 percent, 190 patients aged 1 to 10 died from this condition out of 256. In the 11-20 age range, 22.642 percent of patients died. The whole cumulative percentage is 100 percent.

Ages	*Number*	*Percentage*	*Valid percentage*	*Cumulative percentage*
1 --10	190	71.698%	71.698%	100%
11--20	60	22.642%	22.642%	100%
21--25	15	5.660%	5.660%	100%
TOTAL	265	100%	100%	100%

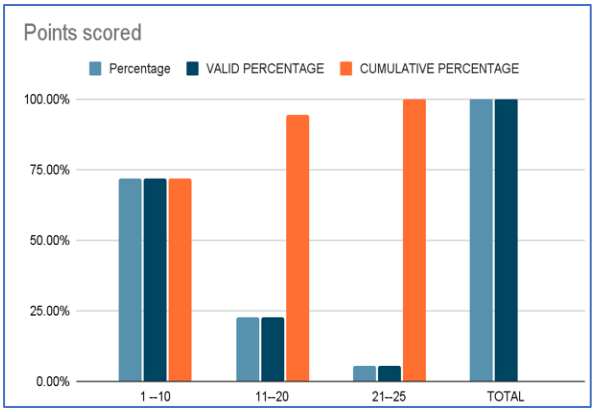


Table 8 displays the death rates of thalassemia patients by gender. With a total of 180 patients and a valid rate of 67.925 percent, the majority of patients are male. while the female number is 85 out of 265 for a rate of 32.075%. 100 percent is the cumulative proportion.

Gender	*Numbers*	*Percent*	*Valid Percentage*	*Cumulative Percentage*
Males	180	67.925%	67.925%	100%
Females	85	32.075%	32.075%	100%
TOTAL	265	100%	100%	100%

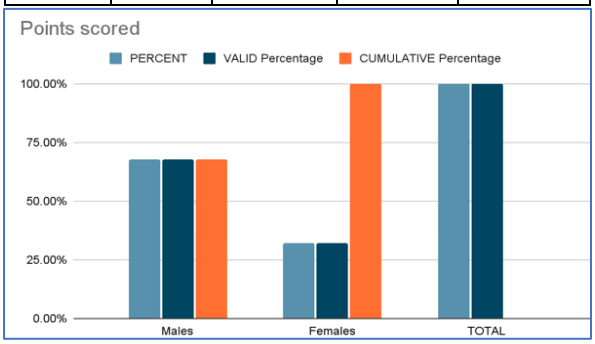
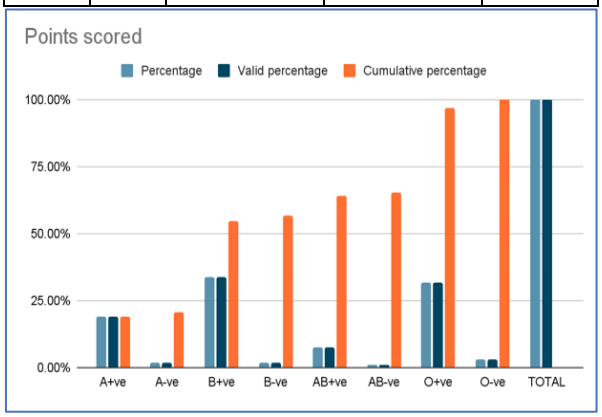


Table 9 displays the death distribution of thalassemia patients by blood type. The majority of patients have the O-positive blood type, as 84 out of 265 patients, or 31.698 percent, are positive. In contrast, 18.868 percent of the 50 individuals with the A+ve blood type died from thalassemia. In addition, the overall percentage is 100 percent.

Blood groups	*Number*	*Percentage*	*Valid percentage*	*Cumulative percentage*
A+ve	50	18.868%	18.868%	100%

A-ve	5	1.887%	1.887%	100%
B+ve	90	33.963%	33.963%	100%
B-ve	5	1.886%	1.886%	100%
AB+ve	20	7.547%	7.547%	100%
AB-ve	3	1.132%	1.1320%	100%
O+ve	84	31.698%	31.698%	100%
O-ve	8	3.0189%	3.0189%	100%
TOTAL	265	100%	100%	100%



Discussion

The purpose of the research was to investigate the causes of Thalassemia major among patients' families in Pakistan, particularly in Sukkur. This study's aims were achieved via the use of questionnaires and discussion based on observation. SPSS was used to examine the thalassemia patient's data. The primary objective is to determine the incidence of thalassemia among various subcastes of the Indian Sindhi community, which has a high frequency of this illness in general. In the Sindhi community of Wardha, prevalence of thalassemia is greater than in previous research and comparable to the most common groups in India. The majority of Sindhis suffer from beta thalassemia, one of the most common forms. It is characterized by reduced synthesis of normal adult hemoglobin (Hb A), the most prevalent form of hemoglobin.

According to our research, 53.96 % in the age range of 1 to 10 experience the greatest suffering. While the rate for male patients is 64.924 %, which is higher than it is for females. Patients with the blood type A+ve have a high prevalence of illness (37.099%). According to our study, couples getting married is the primary cause of thalassemia, accounting for the greatest proportion at 93.255 %.

The major signs and symptoms of thalassemia include swelling. Patients with thalassemia have effects on their physical health, with anxiousness and stress being the two primary symptoms.

Our research indicates that Sindh likewise has a high fatality rate. males had the greatest death rate, or 67.925 % O +ve is a high proportion, at 31.698 percent, while the death rate is high in the 1-10 age group, where it is 71.698 %.

Conclusion

Pakistan is one of the nations with the largest thalassemia prevalence in the world. There are 100,000 thalassemia patients in the nation who need on transfusions, according to a typical statistic. This essay tries to consider the challenges that patients and families with thalassemia in Pakistan are now dealing with. In addition, it offers a futuristic answer by developing a national strategy and strategic plan.

We find that almost half of siblings born to -thalassemia carriers are also thalassemia patients. Additionally, males had a greater incidence of thalassemia, whereas thalassemia patients had higher levels of the blood O +ve blood type.

Recommendations

The research has advised taking action to stop and slow the spread of thalassemia.

• Through print and electronic media, thalassemia should be made more widely known and the public should get health education.

• It is suggested that the thalassemia centers take action to maintain accurate records and cooperate with data gathering. The hospitals and patient welfare organizations must work with the researchers since data gathering is their greatest challenge in Pakistan. Controlling the illness is not only the duty of thalassemia welfare societies and public health institutions. But every person who has thalassemia also has a responsibility, and it is particularly important for carrier moms to see concerned physicians often while they are pregnant.

• Every basic health unit must have all the necessary medical supplies. The government should provide a fair amount in the health budget for thalassemia treatment and prevention.

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Conflicts Of Interest

There are no conflicting interests.

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