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ROLE OF ULTRASOUND IN ASSESSMENT OF SPLEEN IN PATIENTS WITH THALASSEMIA

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Abstract:

Background: Thalassemia is a genetic blood disorder that can lead to significant splenic complications. Ultrasound assessment of the spleen in thalassemia patients is crucial for early detection and management of these complications.

Objective: To sonographically assess spleen characteristics in patients with thalassemia.

Methods: This prospective observational study included 101 thalassemia patients at Dastak hospital, Sabzazar, Lahore, over a 9-month period. Patients of all ages and genders with confirmed thalassemia diagnoses were included, while normal individuals, those with abdominal distension, and previous abdominal surgery were excluded. Ultrasound examinations were performed to assess spleen size, volume, and echogenicity.

Results: The mean age of patients was 7.32 years, with 68.3% having thalassemia major and 31.7% thalassemia minor. The mean spleen size was 13.64 cm, and the mean volume was 83.92 cm^3. Splenomegaly was observed in 65.3% of patients, with 74.3% showing hyperechoic spleens. A significant association was found between patient age and spleen

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abnormality (p = 0.036), as well as between thalassemia type and increased spleen echogenicity (p = 0.001).

Conclusion: Ultrasound assessment revealed significant splenic changes in thalassemia patients, with associations between spleen characteristics, age, and thalassemia type. These findings emphasize the importance of regular sonographic monitoring of the spleen in thalassemia patients for guiding treatment and assessing disease progression.

Keywords: Thalassemia, Ultrasound, Spleen Assessment, Splenomegaly, Hemoglobin Disorders, Spleen Echogenicity.

Introduction:

The name thalassemia derived from a combination of two Greek words: Thalassa meaning the sea [1-3] that is the Mediterranean and anemia ("weak blood"). Another term found in literature, although infrequently, is Cooley's anemia after the name of Prof. Cooley Thomas, a pediatrician in the USA who first described the clinical characteristics of this disorder in patients of Italian origin 1925. [1,2]

Thalassemia is genetic blood disorders inherited from a person's parents that can result in the abnormal formation of hemoglobin [4, 5]. There are two main types, alpha and beta Thalassemia [4]. The severity of alpha and beta thalassemia depends on how many of four genes for alpha or two genes for beta globin are missing [5]. Thalassemia' are widespread throughout the Mediterranean region, Africa, the Middle East, the Indian subcontinent and South-East Asia [6]. As of 2013 thalassemia occurs in about 208 million people with 4.7 million having severe disease [7]. It resulted in 25,000 deaths in 2013 down from 36,000 deaths in 1990 [8]. Males and females have similar rates of disease [9].

Ultrasound assessment of the spleen in thalassemia patients is crucial due to the high prevalence of splenic complications associated with this genetic blood disorder. Splenomegaly, a common condition in thalassemia caused by increased hemolysis and extramedullary hematopoiesis, can lead to hypersplenism, further exacerbating anemia

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and increasing the risk of infections. Regular ultrasound monitoring allows for early detection and tracking of splenic enlargement, facilitating timely medical interventions such as splenectomy when necessary. It also detects splenic infarcts, fibrosis, and vascular changes, which are critical for understanding the overall health of the spleen and guiding treatment decisions. Moreover, there is scanty of data on local population.

Objective:

To sonographically assess spleen in patients with thalassemia.

Materials and Methods:

This prospective observational study will be conducted at Dastak hospital, Sabzazar, Lahore, over a period of 9 months. The study will include 100 participants selected using a non-probability purposive sampling technique. Inclusion criteria encompass patients of all ages and genders with confirmed diagnoses of thalassemia, including beta-thalassemia major, beta-thalassemia intermedia, and other variants. The study will exclude normal individuals, patients with abdominal distension, and those with a history of previous abdominal surgery. This research design allows for the systematic collection and analysis of data from thalassemia patients, providing valuable insights into the condition's characteristics and management strategies

Results:

The study included 101 thalassemia patients, with a slight female predominance (50.5% females, 49.5% males). The mean age of patients was 7.32 years (SE 0.2), ranging from 4 to 10 years. The most common age group was 10-year-olds (25.7%), followed by 4-year-olds (17.8%). Regarding thalassemia type, 68.3% of patients had thalassemia major, while 31.7% had thalassemia minor. Ultrasound examination revealed a mean spleen size of 13.64 cm (SE 0.22) and a mean spleen volume of 83.92 cm^3 (SE 2.60).

Spleen echogenicity on ultrasound showed that 74.3% of patients had hyperechoic spleens, 13.9% had spleens isoechoic to the liver, and 9.9% had mildly hyperechoic spleens compared to the liver. Splenomegaly was diagnosed in 65.3% of patients, while 32.7% had mildly enlarged spleens, and only 1.9% had no splenomegaly. The frequency

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of splenomegaly increased with age, with 24 out of 26 ten-year-olds showing splenomegaly.

Statistical analysis revealed a significant association between patient age and spleen abnormality ($p = 0.036$). Additionally, there was a significant association between thalassemia type and increased spleen echogenicity on ultrasound ($p = 0.001$). These findings suggest that both age and thalassemia type play a role in the development of spleen abnormalities in thalassemia patients

Table 1: Age * Spleen Abnormality Cross tabulation

		Spleen Abnormality			Total
		Mildly enlarged spleen	Nil	Splenomegaly	
Age	4	0	7	11	18
	5	0	4	6	10
	6	1	7	5	12
	7	1	7	5	12
	8	0	2	4	6
	9	0	3	14	17
	10	0	2	24	26
Total		2	32	69	101

Table 2: Association of the patient age with spleen abnormality

Chi-Square Tests

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	Value	Df	Asymptotic Significance (2-sided) p value
Pearson Chi-Square	30.109[a]	18	.036
Likelihood Ratio	29.725	18	.040
N of Valid Cases	101		
a. 19 cells (67.9%) have expected count less than 5. The minimum expected count is .06.			

Thalassemic Patient Age was slightly found to be associated with spleen abnormality as p value was less than 0.05.

Table 3: Type of Thalassemia * Spleen Echogenicity Cross-tabulation

		Spleen Echogenicity				
		Hyperechoic	Hyperechoic To Liver	Isoechoic To Liver	Mild Hyperechoic To Liver	Total
Type of thalassemia	Major	66	0	1	2	69
	Minor	9	2	13	8	32
Total		75	2	14	8	101

Table 4: Type of thalassemia and Spleen Echogenicity on ultrasound

Chi-Square Tests			
	Value	**df**	**Asymptotic Significance (2-sided)**
Pearson Chi-Square	53.189	4	0.001

Thalassemia was significantly associated with increased Spleen echogenicity on ultrasound as p value was less than 0.05

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Discussion:

The study of spleen characteristics in thalassemia patients has been a subject of significant research interest, with various studies contributing to our understanding of splenic changes in this condition. Our findings, when contextualized within the broader literature, provide valuable insights into the sonographic features of the spleen in thalassemia patients.

Our study revealed a mean spleen size of 13.64 cm and a mean spleen volume of 83.92 cm^3 in thalassemia patients. These findings are notably different from those reported by Afridi et al. (2023) [10], who observed a much larger mean splenic volume of 320.62 cm^3 in patients receiving whole blood transfusions. This discrepancy could be attributed to several factors, including differences in transfusion protocols, patient populations, or disease severity. Interestingly, our results align more closely with the findings of Nemati et al.,[11] who established normal ultrasonographic limits for spleen length and volume in healthy Caucasian children. This comparison suggests that our patient population may have had better-controlled disease or more effective management strategies.

The prevalence of splenomegaly in our study (65.3%) is comparable to the findings of Al-Salem et al. (2018)[12], who reported persistent splenomegaly in Saudi thalassemia patients well into adulthood. However, our study showed a lower rate of autosplenectomy (1.9%) compared to Al-Salem's 6.6%. This difference could be due to variations in patient age ranges, disease severity, or management approaches.

Our study revealed a significant association between patient age and spleen abnormalities ($p = 0.036$). This finding is consistent with several other studies in the literature. Al-Salem et al.[12] observed that the splenic index increased with age until about 40 years before gradually decreasing. Similarly, Walker et al. (2022)[13] reported an age-related decline in splenic length in homozygous sickle cell disease patients, which was influenced by genetic factors known to inhibit sickling. The age-related changes in spleen size and function highlight the importance of long-term monitoring in thalassemia patients. As

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Kolnagou et al. (2013)[14] suggested, regular sonographic assessment of the spleen is crucial for evaluating iron overload and guiding chelation therapy.

In our study, 74.3% of patients had hyperechoic spleens. This finding is consistent with the observations of Eze et al. (2015)[15], who reported abnormal spleen parenchyma of varied appearances among thalassemia subjects. The significant association we found between thalassemia type and increased spleen echogenicity ($p = 0.001$) adds to the growing body of evidence suggesting that ultrasound can be a valuable tool in assessing splenic changes in thalassemia patients. Sadeq et al. (2024)[16] further corroborated these findings, noting that increased echogenicity is more frequently seen in patients with small spleens (87.5%, $p<0.001$). This relationship between spleen size and echogenicity underscores the complex nature of splenic changes in thalassemia and the importance of comprehensive sonographic assessment.

The findings from our study and the broader literature have several important clinical implications. First, they underscore the value of ultrasound as a non-invasive, reliable method for assessing splenic changes in thalassemia patients. As Picardi et al. (2012)[17] demonstrated, ultrasound-measured volume was the most sensitive method for identifying non-palpable splenomegaly in patients with primary myeloproliferative diseases. Secondly, the age-related changes in spleen size and function highlight the need for regular, long-term monitoring of thalassemia patients. This is particularly important given the findings of Aessopos et al. (2014) [18], who observed that thalassemia patients with extensive splenomegaly are characterized by high output state and portal hypertension.

Conclusion:

This study highlighted the importance of ultrasound in assessing splenic changes in thalassemia patients, revealing significant associations between spleen characteristics, age, and thalassemia type. The findings suggested the need for regular sonographic monitoring of the spleen in thalassemia patients to guide treatment and assess disease progression

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