Original Article

Health-Related Quality of Life among Iraqi Patients with Systemic Sclerosis

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Abstract

Background: Scleroderma, or systemic sclerosis (SSc), is an extremely rare condition that can have a devastating effect on an individual's physical and mental well-being, as well as a major impact on their ability to go about their everyday lives. Since a cure is currently unavailable, the goal of SSc treatment is to alleviate impairment and symptoms while simultaneously enhancing quality of life.

Objective: The objective is to examine the health and quality of life as a whole in patients diagnosed with systemic sclerosis (SSc) by using the Short Form 36 (SF-36) questionnaire. Additionally, we want to establish a correlation between the SF-36 scores and the patient's clinical and epidemiological features.

Patients and methods: Using the criteria established as per the 2013 ACR/EULAR guidelines, this study utilized a cross-sectional design that included 42 SSc patients. Quality of life was assessed by the SF-36 questionnaire and various demographic, clinical, and epidemiological data were gathered.

Results: Correlations were found between the quality of life scores and age and education level and most domains of clinical characteristic (skin thickening component; fingers, hand, forearm, Puffy fingers, abnormal nail fold capillaries, Raynaud's phenomenon, shortness of breath, oral puckering, interstitial lung disease (ILD)) (P-value < 0.05), interestingly, long duration of disease, had significant impact on quality of life (p-value=0.005).

Conclusion: A decreased QoL is observed among SSc patients. Skin thickening, puffy fingers, telangiectasia, abnormal nail folds, Raynaud syndrome, shortness of breath, oral puckering, and ILD were prevalent in our research.

Keywords: Scleroderma, Systemic sclerosis, quality of life.

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Introduction

Scleroderma, or systemic sclerosis (SSc), is a complex and rare disorder that affects the conective tissues. The pathogenesis of systemic sclerosis (SSc) is marked by the first impairment of tiny blood vessels, inflammation around blood vessels, and aberrant modulation of the immune system. These variables ultimately result in fibrosis in both the skin and internal organs. Myofibroblasts have a significant role in driving the fibrosis process.[1,2] The primary cause of the high death rate in SSc is mostly due to complications affecting internal organs, particularly interstitial lung disease (ILD) and significant cardiac involvement. These complications significantly affect both the length and quality of a patient's life. Scleroderma develops in a chronic manner. There are two distinct forms of SSc: limited cutaneous involvement (LcSSc), which causes thickening of the skin primarily on the extremities and face, and diffuse cutaneous involvement (DcSSc), which affects the whole body.[3] Furthermore, the disease's substantial impact on orofacial functions, everyday activities, and appearance renders it a true social burden.[4] Genetic susceptibility factors play a vital role in

the onset and advancement of SSc, but the precise etiology of the disease remains unknown.[5] Presently, the management of skin abnormalities or lesions in SSc primarily aims to alleviate symptoms and restore organ function, presenting a considerable obstacle for healthcare practitioners.[4] Evaluating the quality of life is essential for the treatment of the condition and may provide useful insights for identifying appropriate medicines. These therapies will likely be administered by a multidisciplinary team, and are expected to be non-pharmacological in nature.[6] The emphasis on enhancing the mental and physical health of persons with scleroderma stems from a holistic approach to managing patients with chronic, inflammatory, and connective tissue illnesses, which is fundamental to the field of rheumatology medical care. The first Asian research examined the comparative health-related quality of life (HRQoL) of individuals with autoimmune illnesses. The findings revealed that patients with systemic sclerosis (SSc) had more significant impairments in mental health and had a worse sense of overall health. Furthermore, the study found that the severity of skin involvement in SSc patients correlated to lower physical and mental QoL scores. Patients with SSc reported a much worse health-related quality of life than healthy individuals.[7] As far as we know, Among Iraqi populations, this is the first study that focuses on the clinical-epidemiological aspects of SSc and its effects on quality of life.

Methods

This cross sectional study was conducted from April 2023 to October 2023 at Baghdad Teaching Hospital's Rheumatology Unit, Department of Medicine. Examining the correlation bet-ween clinical-epidemiology variables and quality of life, it surveyed Iraqi patients diagnosed with systemic scleroderma. All individuals who took part in the research gave their informed per-mission after receiving ethical clearance from the medical department at Baghdad Teaching Hospital and the College of Health and Medical Techniques in Baghdad. And the research followed all of the guidelines laid out by the 1964 Declaration of Helsinki when it involved human subjects.

During the study, a group of forty-two adult patients participated. The patients were diagnosed with scleroderma in accordance with the categorization criteria for systemic sclerosis that were established by the American College of Rheumatology and the European League Against Rheumatism in 2013. We excluded individuals who satisfied any of the following criteria: inability to completely engage in the study due to their medical condition, a previous diagnosis of mental illness, cancer, another chronic disease, unwillingness to cooperate, or handicap.

The Short-Form Health Survey Score Questionnaire 36 (SF-36) used the validated Arabic version. The questionnaire is a self-administered tool to assess the quality of life and health. It comprises two primary components, namely physical and mental component scores (PCS and MCS, respectively). Within these components are eight main scales, each comprising 36 questions. These scales cover various aspects such as general health, physical functioning, physical role, pain, vitality, social functioning, emotional problems, and mental health. The score for each scale varies from zero to one hundred. A higher score suggests an improved health-related quality of life.[8]

Results

An excel document included encoded questionnaire replies. Data was analyzed using IBM SPSS-29 (IBM Statistical Packages for Social Sciences, version 29, Chicago, IL, USA). Simple frequency, percentage, mean, standard deviation, and range measurements were used.

Students-t-test, Paired-t-test, or ANOVA tests were used to determine the significance of difference of means (quantitative data). The significance of percentage differences in qualitative data was assessed using Pearson Chi-square test (χ^2-test) with Yate's correction or Fisher Exact test where appropriate. P values of 0.05 or less indicated statistical significance.

Table (1) displays the correlation between the quality of life that assessed by the SF-36 questionnaire score for the analyzed sample and their

socio-demographic and epidemiological features. The table identified a statistically significant correlation between age and education level (p-value > 0.05). Additionally, based on epidemiological characteristics, a statistically significant association was found between the disease subtype and the presence of the disease (p-value = 0.005). Furthermore, it was observed that a longer duration of the disease significantly impacted the QoL (p-value=0.005).

Table (2) demonstrates the association between disease impacts on quality of life assessed by SF-36 questionnaire score and clinical features. A significant association was found between the SF-36 questionnaire score and most domains of clinical characteristics, include (skin thickening component; fingers, hand, forearm, Puffy fingers, telangiectasia, abnormal nail fold capillaries, Raynaud's phenomenon, shortness of breath, oral puckering, ILD, infection) (P-value < 0.05).

Table 1: *Association between quality of life score and sociodemographic and epidemiological characteristics.*

Sociodemographic Characteristics.		**SF-36 Questionnaire Score**		
		No.	**Mean± SD**	**P value**
Age (years)	< 30 years	6	54.1±27.5	0.008**
	30 — 39	9	26.0±16.3	
	40 —49	12	24.0±15.1	
	50 —59	10	24.3±16.1	
	≥ 60 years	5	20.6±3.9	
Sex	Male	3	39.7±18.7	0.301
	Female	39	27.5±19.5	
Residence	Urban	34	30.3±20.1	0.182
	Rural	8	20.1±14.2	
Education level	Illiterate	19	21.6±12.2	0.009**
	Primary	10	33.9±21.6	
	Intermediate	5	23.1±9.0	
	Secondary	5	53.4±28.8	
	College	3	20.6±13.5	
Occupation	Employed	2	14.6±10.0	0.309
	Unemployed	40	29.1±19.6	
Marital status	Single	7	27.6±16.4	0.160
	Married	31	30.8±20.3	
	Divorced	4	11.0±5.5	
BMI (Kg/m^2)	Underweight (<18.5)	11	22.7±12.8	0.682
	Normal (18.5-24.9)	14	28.8±22.0	
	Overweight (25-29.9)	11	33.0±22.7	
	Obese (≥ 30)	6	29.4±19.1	
Epidemiologic Characteristics		**SF-36 Questionnaire Score**		
		No.	**Mean±SD**	**P value**
Interval between onset of disease & diagnosis/years	< 1 year	13	35.6±23.1	0.306
	1 — 4	16	28.0±19.1	
	5---9	7	24.8±17.8	
	= > 10 years	6	18.1±8.1	
Duration of disease/years	< 1 year	-	-	0.005**
	1 — 4	10	44.9±27.2	
	5 — 9	15	25.3±14.7	
	= > 1 0years	17	21.3±11.5	
Disease subtype	Localized	3	57.7±44.0	0.005*
	Systemic	39	26.1±15.3	
Systemic subtype	Limited	12	24.6±19.3	0.688
	Diffuse	27	26.8±13.4	

*Significant difference between two independent means using Students-t-test at 0.05 level.
**Significant difference among more than two independent means using ANOVA-test at 0.05 level.

Table 2: *Association between quality of life score and clinical features.*

Clinical Features		SF-36 Questionnaire Score		
		No.	Mean±SD	P value
Signs Symptoms of Scleroderma				
Skin thickening				
Fingers	Yes	39	26.1±15.3	0.005*
	No	3	57.7±44.0	
Hand	Yes	39	26.1±15.3	0.005*
	No	3	57.7±44.0	
Forearm	Yes	37	25.5±14.1	0.008*
	No	5	49.4±38.1	
Trunk	Yes	27	26.8±13.4	0.484
	No	15	31.2±27.5	
Puffy fingers	Yes	27	23.6±13.6	0.031*
	No	15	37.0±25.3	
Digital ulcer	Yes	22	23.1±14.8	0.064
	No	20	34.2±22.5	
Fingertip pitting scars	Yes	24	25.4±16.6	0.259
	No	18	32.3±22.6	
Telangiectasia	Yes	35	25.8±14.6	0.048*
	No	7	41.6±33.7	
Abnormal nail fold capillaries	Yes	37	25.5±15.2	0.007*
	No	5	49.7±33.9	
Raynaud's phenomenon	Yes	38	25.9±15.4	0.009*
	No	4	51.9±37.8	
Dysphagia	Yes	31	25.6±12.6	0.114
	No	11	36.4±31.3	
Shortness of breath	Yes	30	22.7±11.5	0.002*
	No	12	42.6±27.4	
Diarrhea	Yes	17	25.6±13.5	0.455
	No	25	30.3±22.7	
Weight loss	Yes	18	26.1±14.5	0.516
	No	24	30.1±22.6	
Sclerodactaly	Yes	30	24.7±12.4	0.051
	No	12	37.6±29.6	
Oral puckering	Yes	31	23.9±12.8	0.011*
	No	11	41.0±28.7	
Skin lesion	Yes	3	57.7±44.0	-
	No	-	-	
Complication of scleroderma				
Pulmonary hypertension	Yes	8	19.0±11.9	0.132
	No	34	30.6±20.3	
Interstitial lung disease	Yes	31	24.5±13.4	0.027*
	No	11	39.4±28.8	
Gangrene	Yes	1	7.1±	0.273
	No	41	28.9±19.4	
Myositis	Yes	1	4.9±	0.225
	No	41	29.0±19.3	
Infection	Yes	9	15.1±8.9	0.019*
	No	33	32.0±20.0	
Renal failure	Yes	3	11.7±7.5	0.125
	No	39	29.7±19.5	

*Significant difference between two independent means using Students-t-test at 0.05 level.
**Significant difference among more than two independent means using ANOVA-test at 0.05 level.

Table 3. Showed that the SF-36 total mean score was (28.39), total mean score on (PCS) was 28.17 and in (MCS) was 28.64.Moreover all scores in the table showed the poor score was in role of

physical health problems and emotional health items. While the greater score was in social functioning (49.7).

Regarding the overall SF-36 questionnaire score of the studied sample about all items of score, only 14.3% of the studied sample had a good score, while the majority (54.8%) had a poor score, and 31.0% had fair and acceptable scores.

Table 3: *SF-36 mean Score stratified for disease characteristics.*

SF-36 Scale	Mean Score
physical component summary scores (PCS)	28.17
General Health	36.46
Physical function	30.83
Role limitation – physical	11.90
Bodily pain	33.46
Mental component summary scores (MCS)	28.64
Role limitation – emotional	12.70
Social functioning	49.7
Vitality	24.05
Mental health	28.1
Total SF-36 Score	28.39
*Values was expressed as mean.	

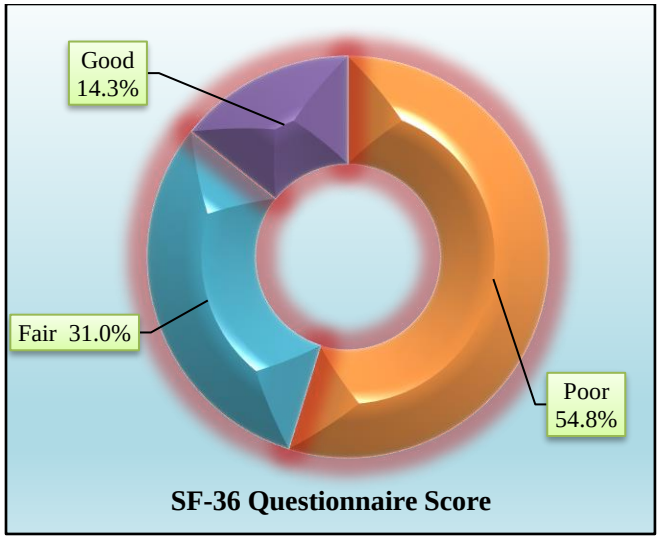

Figure 1: *Total score of the scleroderma patients assessed by SF-36 questionnaire score.*

Discussion

Heterogeneous connective tissue diseases including fibrosis, vasculopathy, and immunological activation are known as systemic sclerosis (SSc). A patient's QoL is negatively impacted by the disease's multisystem involvement, which has serious psychological and physiological effects. Quality of life (QoL) is a multi-faceted concept that depends on people's perceptions of their health and how those perceptions interact with other, less health-specific parts of their lives.

There is a scarcity of data about the characteristics of patients with scleroderma from the Middle East. Our research indicates that this is the first cross-sectional research in Iraq that looks at the relationship between clinical and epidemiological factors and the effect of scleroderma on patients' quality of life, whether considering all cases of scleroderma or dividing it into two subtypes: systemic and localized. This research provides new insight on the correlation between the severity of SSc and HRQoL; it was carried out in Iraqi patients.

The average age was 44.6 years, with an SD of 12.7. Based on other research in various countries, these findings confirm that systemic sclerosis typically manifests in individuals during their fourth decade of life. This pattern has been observed in various countries, such as Iraq[9] where the average age was 40±11 years;[10] in Libya, with an average age of 40.5±21.5 years; in Egypt[11] where it was reported as 44.50 ±11.17 years, and; in Iran where the mean age was 44 years.[12] The present study disagrees with several studies that reported older respondents, such as[13] in Thailand (61.3±9.9 years), in Milan, Italy (56.6 ± 13.3 years)[14] and in Turkey (50.9±12.4 years).[15] In addition, there were other studies that revealed lower average ages than the one we used. For example, Pakistan[16] reported 34.5±1.5 years as the average age, Bangladesh[17] 36.5±11.3 years, and another Iraqi research18 was 36.8 ± 11.4 years. Possible causes of the differences include research design, patient selection, environmental factors, and genetics.

Among the many interesting aspects of this research, it appears that the female preponderance was high, with an even larger female-to-male ratio of 13:1 within a percentage of 92.9%; and 7.1% were men. Furthermore, these findings were in line with prior research. In Libya, Elbraky et al,[10] analyzed the findings of the 30 patients, which

consisted of 28 females (representing 93.3% of the total) with a female-to-male ratio of 14:1); Asif et al,[16] a Pakistani study showed a higher percentage of females compared to males with a ratio 16:3, from total 38 patient female percentage was 84.2%.

In the current study, 39 patients (92.9%) had Systemic scleroderma, three patients (7.1%) had Localized scleroderma**,** according to systemic subtype, finding 12 patients (30.8%) had limited systemic sclerosis, and 27 patients (69.2%) had diffused SSc.

In contrast to the literature, which indicates a higher percentage of patients suffering from limited subtypes, this research found a higher percentage of patients with diffuse scleroderma. Such as Gorial & Swady[18] found (Limited 60.0% vs Diffuse 40.0%)**.** In Poland, Żebryk et al,[19] revealed (Limited 56% vs Diffuse 33%)**.** The current results align with the published data from many places, as shown in Elbraky et al,[10] who reported in their study that (80%) had DcSSc and (20%) had LcSSc in Libyan patients, Nigeria (57.1%), Qatar (52.4%).[10] In UAE Overall, (Limited 45.5% vs Diffuse 54.5%).[20] In Egypt, (Limited 28% vs. Diffuse72%).[21]

The analysis of the existing findings based on clinical manifestations revealed that the present results highlight a significant association between scleroderma types and QoL. Furthermore, the health problems were more serious in those with DcSSc, leading to a worse quality of life compared to those with LcSSc. However, this discovery is interesting; the analysis could not demonstrate that the systemic sclerosis subtype is an independent factor that influences quality of life. Therefore, it is recommended that therapies that focus on enhancing QoL be equally focused on both disease categories. This finding is similar to Sierakowska et al,[22] which showed that disease type is not significantly associated with quality of life. Moreover, Alian et al,[21] has revealed that individuals with DcSSc have more extensive organ involvement compared to those with a limited subtype. While, Sherby et al in 2022 reported that no significant correlation was seen between disease subtypes with the quality of life (QoL) of patients. However, the findings do not differentiate between the disease subtypes, and there was no statistically significant difference in terms of quality of life or SF-36 survey scores between the two groups.

The current study demonstrated a correlation between longer illness duration and related to DcSSc patient subgroup with a decrease in QoL. This confirms what Korean research found: a worse physical quality of life was correlated with a longer duration of illness, involvement of the gastrointestinal tract and lungs, and mRSS. These findings corroborate what has been previously reported.[22]

On the other hand, some studies are not consistent with current findings. For example, Sherby et al,[23] revealed there is no statistical clinical correlation between the length of illness and the quality of life in people with SSc. Allanore et al,[24] revealed no significant associations between disease duration and Quality of life score in their research. Chevreul et al,[25] documented that's no association between HRQoL and disease duration. This difference may be due to using different QoL assessment tools, more complications, or more medication use.

This study revealed that SSc has various impacts on patients' physical health, mental health, and overall quality of life (QoL), including (skin thickening, puffy fingers, telangiectasia, abnormal nail fold, Raynaud phenomenon, shortness of breath, oral puckering, ILD, Infection). One of the primary indications of systemic sclerosis (SSc) is the Raynaud phenomenon, and these results also follow several studies demonstrating that the Raynaud Phenomenon significantly impacts the QoL in Systemic Sclerosis, making it one of the most prevalent symptoms, such as Allanore et al,[24] indicated significant correlations between QoL and organ involvement, including the lungs, heart, Raynaud Phenomenon, digestive system, kidneys, and skin. Hossain et al,[17] also found similar findings, supporting the idea that Raynaud Phenomenon considerably impairs the quality of life of the patients in our

research. According to Van Leeuwen et al,[26] Raynaud Phenomenon is a prevalent symptom that significantly affects the QoL of patients with SSc. Pauling et al,[27] found that the predominant symptom in their research is Raynaud's phenomenon in patients with SSc, which significantly reduces QoL. Raynaud's illness, along with the adverse effects of internal organs, particularly the lungs, could complicate the performance of daily activities as well as restrict social functioning.[28] In disagreement with the present study, Sherby et al,[23] found that Raynaud's phenomenon and gastrointestinal symptoms were more frequent in patients. This manifestation wasn't statistically associated with poor quality of life in the examined cohort. According to the SSc SF-36 score, other research suggests these manifestations may pose problems for SSc patients. Skin involvement was also linked to poor physical and mental QoL in SSc patients.[7] Van Leeuwen et al,[26] indicate that hand function deficits, as measured by finger-to-palm distance and grip strength, significantly affect QoL with time. Sierakowska et al,[22] showed that health problems (ulceration of the fingers, disability, and dyspnea) were more severe in the DcSSc, and consequently, the quality of life in patients with DcSSc was worse than those with LcSSc. Additionally, musculoskeletal and skin involvement is strongly linked to a worse quality of life.[17] In their multivariable study, Van Leeuwen et al,[26] ILD did not impact HRQoL with time, contrary to predictions. This may be because not all SSc-ILD patients have ILD symptoms. In a retrospective investigation of 378 SSc patients, Ciaffi et al, found 156 with ILD and pulmonary arterial hypertension. Lung function alterations significantly affect HRQoL.[29]

Clinical signs like pulmonary hypertension have already been described as a predictor of poorer quality of life in current SSc patients, but these associations were not identified in current and Brazilian populations.[30] It's possible that the sample size wasn't big enough to find these correlations, even though the sickness may have different effects on different groups of people.

Extracellular matrix components, such as collagen, are increased in SSc patients, which causes gastric difficulties, in the upper and lower GIT. Gastroesophageal reflux is one of the leading causes of gastrointestinal discomfort.[17] On the other hand, the current patients' gastrointestinal problems had little to no effect on their day-to-day functioning or overall well-being; this agrees with Hossain et al.[17] GIT symptoms in SSc patients had a minor impact on quality of life, according to Läubli et al.[31] Patient quality of life are most affected by SSc, Raynaud symptoms, digital ulcers, and gastrointestinal problems. Over time, persistent pulmonary arterial hypertension lowers health-related quality of life.[26]

Patients suffering from gastrointestinal symptoms had a worse quality of life compared to those who did not, according to research by Sherby et al.[23] This finding is in line with that of van Leeuwen et al,[26] who also found that gastrointestinal problems negatively impact SSc-related QoL. Different techniques used to measure quality of life could explain these discrepancies.

This research showed that, On the SF-36, out of the eight domains, the one with the greatest score was social functioning, while the one with the lowest score was role limitation—physical and emotional role. Within the total SF-36 score of 28.39±19.44, PCS mean score of 28.17, and MCS mean score of 28.64, this result is in line with the research conducted by Hossain et al,[17] in Bangladesh, where they observed that the Mental Component Summary (MCS) score (42.0) was greater than the Physical Component Summary (PCS) score (26.2). This suggests that the mental aspect of the SF-36 questionnaire was better compared to the physical aspect. In contrast, in Egypt, Sherby et al,[23] found total mean SSc QoL score 20.2 ± 6.5 which agree with the current result. On the other hand, March et al,[32] reported SF-36 total score 54.6±17.2, and Hudson et al. Hudson et al, in 2009 report PCS score 36.7 (11.2) and MCS score 49.0 (11.7). In a recent study, the findings of Van Leeuwen et al,[26] revealed average SF-36 scores for the MCS were 62.6 (SD 22), whereas average scores in the PCS were 47.7 (SD 20). In accordance with the present finding, Hossain et al,[17] explain that SSc patients adapted well to their gradual progression of disease, and their study also found

that the MCS score was greater than the PCS score, suggesting that the mental aspect of SF-36 was better than the physical aspect.

In this research, the categories of social functioning had the highest SF-36 scores, whereas the categories of role limitation—physical and emotional—had the lowest scores. The greatest score was for social functioning, according to the Egyptian research.[21] In addition, compared to the physical and emotional role domains, the social functioning domain had the greatest score, as shown by Bretterklieber et al,[34] and Hossain et al,[17] Also, consists with Rosso et al,[35] discovered that the emotional domain performed better than the others. The mental health domain also obtained the best result compared to the other domains, according to Racine.[36]

Previous research supports the current findings. An Italian study by Danieli et al,[37] utilizing SF36 found that SSc patients' low HRQoL was caused by daily activity restrictions. Sierakowska et al,[22] similarly found that functional impairment affects SSc quality of life.

Several constraints are worth mentioning. The research acknowledges the limited number of patients, the rarity of the diseases, and the fact that it was conducted at a single facility. However, it is important to highlight that the sample size is similar to that of numerous previous studies. Expanding the sample size and conducting longitudinal research might reveal other interest-ing variations among the reported parameters in this study.

Conclusion

Systemic sclerosis (SSc) patients experience poor quality of life. The SF-36 scores showed that SSc patients with higher social functioning scores were better able to respond to health-related quality of life modifications. Higher SF-36 scores were associated with Skin thickening, puffy fingers, telangiectasia, abnormal nail folds, Raynaud syndrome, and shortness of breath, oral puckering, and ILD, indicating that systemic sclerosis (SSc) patients with more severe illnesses were more resilient to health-related quality of life modifications.

Ethical Approval: The study was approved by the Ethical Committee of Baghdad Teaching Hospital, Iraq vide IRB number 15357.

Conflict of Interest: There was no conflict of interest to be declared by any author.

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Author's Contribution

AHAH: Conception and design, drafting of article, final approval, critical revision.

DAD: Conception and design, analysis and interpretations, critical revision, drafting of article, final approval.

IAJ: Conception and design, drafting of article.

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