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Period prevalence of Systemic Sclerosis (Morphoea) in Tertiary Care Hospital in India: An Update

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ABSTRACT

Systemic sclerosis (SSc) is a multisystem autoimmune disease characterized by fibrosis and vasculopathy. It is a rapidly progressing disease and wide spreading dysfunctioning of various organs includes kidney, gastro intestinal, cardiovascular systems and etc. Annual incidence is 19 per million, and prevalence is 19-75 per 100,000, with a female: male ratio of 3:1, and 8:1 in mid to late childbearing years. Incidence is twice as high among African Americans. The data was collected from in-patient record book in the Deportment of Dermatology in a tertiary care hospital in India for a period of March 2011 to February 2013.

A total of 180 patients were recorded in the in-patient record book of the Department of Dermatology in a tertiary care hospital for a period of March 2011to February 2013. Among the patients of systemic Sclerosis the dominating age groups were 21 to 30 years and 31to40 years. In the study most of the females 144 (80%) patients out of 180, males 36 (20%) patients out of 180 are having Systemic sclerosis disease. The female/male ratio of disease is 4:1.

Keywords: Systemic Sclerosis, Auto immune disorder, Epidemiology, Prevalence in a tertiary care hospital, Female/Male ratios.

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INTRODUCTION

Systemic Sclerosis (SSc) is a connective tissue disorder in which vascular alterations and endothelial damage are prominent and lead to progressive and widespread dysfunction of various organs. Vascular symptoms such as Reynaud's phenomenon, digital ulcers and pulmonary arterial hypertension are also a frequent target of diagnostic and therapeutic efforts.[1] The localized form of scleroderma, also known as morphoea. This heterogeneity of clinical manifestations in Systemic sclerosis (SSc) has led to efforts to find markers that enable identification of patients most at risk of involvement of particular organ system, who would benefit from more frequent and organ-specific monitoring.[2]

Systemic sclerosis (SSc) is mainly classified into two types, according to the extent of skin involvement.

Limited scleroderma or Limited cutaneous Systemic Sclerosis (LcSSc):

70% cases of systemic sclerosis are LcSSc type. Face, forearms and lower legs up to the knee are mainly affected parts in body. Usually a milder disease, with less skin involvement, slows onset and slow development. The slow onset may mean that symptoms are moderately unobserved until internal complications takes place.[3]

Diffuse scleroderma or Diffuse cutaneous systemic sclerosis (DcSSc):

30% cases of systemic sclerosis are DcSSc type. Generally a more rapid commencement, the skin changes may extend quickly, within a few months of disease onset. Skin changes can remit later than a number of years, with softening of the skin and major development in mobility. Upper arms, thighs or trunk are mainly affected. Internal organ involvement is more naturally. [4]

Sign and symptoms of systemic sclerosis:

- Fatigue, Weight loss.
- Swelling (non-pitting oedema) of fingers and toes.
- Swelling and sclerosis reduce hand movements,-the 'prayer sign'.
- Reynaud's phenomenon.
- Calcinosis- nodules or lumps of chalky material which may break through the skin.
- Tight lips, Telangiectasia.
- Hyper pigmentation and hypo pigmentation.
- Joint pain and swelling.
- Myalgia (due to inflammatory myopathy).[5, 6]

Diagnosis:

- Skin biopsy - may aid diagnosis.

- Hand X-ray may show Calcinosis.
- Nail fold capillaroscopy - helps to assess the likelihood of scleroderma in patients with Reynaud's phenomenon or swollen fingers.
- Thermography with cold confronts helps to evaluate the severity of Reynaud's phenomenon.[4, 6]

Management:

The major mortality and much of the morbidity of Systemic Sclerosis (SSc) arises through the development of specific complications of the disease including organ based complications such as cardiopulmonary, renal or gastrointestinal manifestations. The frequency and diverse nature of these complications makes systematic assessment and long term follow up essential to good management of SSc. In addition it is essential that accurate baseline assessment of each case is undertaken to define the extent and pattern of disease and to ascertain the likelihood of progression and risk of severe life-threatening complications.[7]

Epidemiology:

Annual incidence is 19 per million, and prevalence is 19-75 per 100,000, with a female: male ratio of 3:1, and 8:1 in mid to late childbearing years. Incidence is twice as high among African Americans, and the Choctow Native Americans in Oklahoma have the highest prevalence in the world (469/100,000). There is some hereditary association; some suggestion of immune reaction (molecular mimicry) to a virus, and some cases caused by toxins.[8] Systemic scleroderma is a rare disease with an annual incidence of 1 to 2 per 100,000 individuals in the United States. The interval of peak onset starts at age 30 to 35 and ends at age 50 to 55.[9,10] In the United States, the prevalence of systemic scleroderma is about 50,000 with different studies giving different estimates, usually ranging between 40,000 and 165,000.[10]

Previously reported incidence and prevalence estimates vary greatly according to geographic location and methods of case ascertainment. Classification criteria were not developed until 1980 when the American Rheumatism Association (now the American College of Rheumatology, ACR) proposed criteria to distinguish SSc from other connective tissue diseases.[11]

Monaco *et al.*[12]reported a retrospective review of Italian-origin patients (age ≥16 years) with a diagnosis of SSc criteria based on the ACR (American College of Rheumatology) 1980 classification criteria and the revised LeRoy and Medsger 2001 criteria. Patients were identified from outpatient clinics in the Ferrara district (study area of 346 000 population) from 1999 through 2007. The authors found 118 patients who met the LeRoy–Medsger criteria and 88 patients who met the stricter 1980 ACR criteria. Using the LeRoy–Medsger criteria, the annual

incidence rate and prevalence estimate are, respectively, 43 and 341 cases per million. However, fewer met ACR classification criteria, leading to incidence and prevalence estimates of 32 and 254 cases per million, respectively. They further defined the cases into SSc subsets according to the LeRoy and Medsger system. They had 20 limited SSc and 76 limited cutaneous SSc and 22 diffuse cutaneous SSc. They noted a female predominance with a female to male ratio of 9.7: 1.[12] Another article by Rosa *et al.*[13] examined SSc incidence and prevalence in the city of Buenos Aires, Argentina. They reviewed members of a prepaid health maintenance program between 1999 and 2004 with a total of 98 642 persons. The population was largely white and of European descent with only 3% from other ethnicities. They also used the 1980ACRclassificationcriteria and/or the LeRoy and Medsger criteria to define cases. They reported an annual incidence of 21.1 cases per million and a prevalence of 296 cases per million. They also further divided the cases into diffuse and limited SSc. The incidence and prevalence for diffuse SSc were 6.1 and 57 cases per million, respectively, and for limited SSc were 15.2 and 240 cases per million. This group was the first to report the SSc incidence rate and prevalence estimate in Latin America. Their figures are similar to those reported for the US population as well as for Spain and Australia.[13]

In Taiwan, Kuo *et al.* [14] reviewed cases from the catastrophic illness registry of the Taiwan National Health Insurance Research Dataset and the National Death Registry of Taiwan to estimate the incidence, prevalence and mortality of SSc. A total of 1479 persons with SSc were identified in this study. The annual incidence rate was 10.9 cases per million and the prevalence was 56.3 cases per million.[14]

In north India, Minz *et al.* [15] reported the prevalence of antinuclear antibody (ANA) positive autoimmune disorders. They reviewed ANA-positive files from 1996 to 2006 and did a retrospective analysis of the cases, including both adults and children. They reported a prevalence of SSc at 120 cases per million. They included another subgroup of cases as CTD overlap as 70 cases per million and Reynaud's disease as 50 cases per million.[15]

The data was collected from in-patient record book in the Deportment of Dermatology in a tertiary care hospital in India (Gauhati Medical College and Hospital) for a period of 2 years (March 2011to February 2013).

RESULTS AND DISCUSSION

A total of 180(100%) patients were recorded in the in-patient record book of the Department of dermatology in a tertiary care hospital for a period of 2 years (March 2011to February 2013).

Among the patients, 80(44.4%) patients had systemic sclerosis for a period of March 2011 to February 2012, 100 (55.6%) patients had systemic sclerosis for a period of March 2012 to February 2013 (Table 1).

Table 1: The no of Systemic Sclerosis cases reported in March 2011- February 2013.

Age group (years)	Mar 2011 to Feb 2012		Mar 2012 to Feb 2013	
	Male	Female	Male	Female
UPTO 10yr	0	0	0	0
11yr-20yr	3	12	4	13
21yr-30yr	3	22	5	22
31yr-40yr	3	18	3	18
41yr-50yr	6	10	2	15
51yr-60yr	1	2	6	12
61yr-70yr	0	0	0	0
71yr-80yr	0	0	0	0
TOTAL	16	64	20	80
	16+64 = 80 (44.4%)		20+80 = 100 (55.6%)	
			80+100 = 180 (100%)	

Among the patients of Systemic Sclerosis 80 (44.4%), 15 (18.5%) patients were in the age group of 11-20 years, 25 (31.25%) patients were in the age group of 21-30 years, 21 (26.25%) patients were in the age group of 31-40 years, 16(20.0%) patients were in the age group of 41-50 years, 3 (3.75%) patients were in the age group of 51-60 years were recorded for a period of March 2011 to February 2012. (Table 2)

Table 2: Age and sex distribution of Systemic Sclerosis patients from March 2011 to February 2012.

Age group (years)	Mar 2011 to Feb 2012		Male + female	Male/female sex ratio
	Male	Female		
UPTO 10yr	0 (0%)	0 (0%)	0 (0%)	0:0
11yr-20yr	3 (3.75%)	12 (15%)	15 (18.5%)	1:4
21yr-30yr	3 (3.75%)	22 (27.5%)	25(31.25%)	1:7.33
31yr-40yr	3 (3.75%)	18 (22.5%)	21 (26.25%)	1:6
41yr-50yr	6 (7.5%)	10 (12.5%)	16 (20.0%)	1:1.6
51yr-60yr	1 (1.25%)	2 (2.5%)	3 (3.75%)	1:2
61yr-70yr	0 (0%)	0 (0%)	0 (0%)	0:0
71yr-80yr	0 (0%)	0 (0%)	0 (0%)	0:0
TOTAL	16 (20%)	64 (80%)	16+64 = 80 (100%)	

Among the patients of Systemic Sclerosis 100(55.6%), 17 (16%) patients was in the age group of 11-20 years, 27(27%) patients were in the age group of 21-30 years, 21(21%) patients were in the age group of 31-40 years, 17 (17%) patients were in the age group of 41-50 years, 18(18%) patients were in the age group of 51-60 years were recorded for a period of March 2012 to February 2013. (Table 3)

Among the patients of Systemic Sclerosis, male female ratio was 1:4 in the age group of 11-20 years; male female ratio was 1:7.33 in the age group of 21-30 years; male-female ratio was 1:6 in the age group of 31-40 years and 1:1.6 in the age group of 41- 50 years for a period of March 2011 to February 2012. (Table 2) Among the patients of Systemic Sclerosis, male female ratio was 1:3.2 in the age group of 11-20 years; male-female ratio was 1:4.4 in the age group of 21-30 years, male- female ratio was 1:6 in the age group of 31-40 years and 1:7.5 in the age group of 41- 50 years for a period of March 2012 to February 2013. (Table 3)

Among the patients of systemic Sclerosis the dominating age groups were 21 to 30 years and 31to40 years.

Table 3: Age and sex distribution of Systemic Sclerosis patients from March 2012 to February 2013.

Age group (years)	Mar 2012 to feb 2013		Male + female	Male/female sex ratio
	Male	Female		
UPTO 10yr	0 (0%)	0 (0%)	0 (0%)	0:0
11yr-20yr	4 (4%)	13 (13%)	17 (17%)	1:3.2
21yr-30yr	5 (5%)	22 (22%)	27 (27%)	1:4.4
31yr-40yr	3 (3%)	18 (18%)	21 (21%)	1:6
41yr-50yr	2 (2%)	15 (15%)	17 (17%)	1:7.5
51yr-60yr	6 (1%)	12 (12%)	18 (18%)	1:2
61yr-70yr	0 (0%)	0 (0%)	0 (0%)	0:0
71yr-80yr	0 (0%)	0 (0%)	0 (0%)	0:0
TOTAL	20 (20%)	80 (80%)	20+80 = 100 (100%)	

CONCLUSION:

The study showed that among the 180 patients 80 patients were having Systemic Sclerosis for a period of March 2011 to February2012; 100 patients were having Systemic sclerosis for a period of March 2012 to February 2013, thus result shows that prevalence of Systemic Sclerosis increase by year to year. In our study, the patients in the age group of 21- 30and 31-40 are mainly having Systemic Sclerosis. In the study most of the females 144 (80%) patients out of 180, males 36 (20%) patients out of 180 are having Systemic sclerosis disease. The female/male ratio of disease is 4:1.

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