#220 Patient Reported Burden of Indolent Systemic Mastocytosis in White vs. Non-White Patients

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SCAN ME

Introduction

- Systemic mastocytosis (SM) is a rare, clonal mast cell disease driven by the *KIT* D816V mutation. Among its various subtypes, indolent SM (ISM) is the most common
- Because it is a rare disease and presents with heterogeneous symptoms, SM is often characterized by misclassification and substantial delay in accurate diagnosis. These challenges may differ by race or ethnicity

Objectives

- To better understand and explore the potential racial disparities in the ISM patient journey and patient-reported burden of disease following diagnosis, especially related to symptoms, medical resource use, quality of life, and employment

Methods

- Adult patients ≥18 years of age with physician-confirmed ISM according to WHO 2016 criteria were invited to participate in a survey. Patient survey data was supplemented with administrative data extracted from the Research Data Warehouse of Kaiser Permanente Southern California (KPSC)
- The patient survey, including investigator-developed questions and the questions of 3 validated instruments (ISM-SAF, PGIS, and SF12v1), was administered Oct 2023 - Jan 2024
- Demographics, medical resource use, and patient-reported symptoms, quality of life, and work status were assessed
- The patient self-reported disease burden between White and Non-white patients were compared using Kruskal-Wallis and Fisher's exact tests

Results

- 51 eligible ISM patients were invited to participate, 40 completed the survey (response rate = 78.4%). Survey respondents exhibited similar demographics to those who did not respond except for higher family income and education
- Survey response rate for White patients was 100% (25/25) and for Non-white patients was 57.7% (15/26)
- 28% of all ISM patients reported that their SM condition had impacted their ability to work and 28% reported reduced hours at work; 10% had gone on medical disability due to ISM. Ability to work was impacted in 32% of White ISM patients and 20% of Non-white patients

Table 1. Patient Demographics

	Race/Ethnicity Group		
	Non-White (N = 15)	White (N = 25)	Total (N = 40)
Age (years)			
At diagnosis	50.5±12.1	50.7±14.0	50.7±13.1
Range at diagnosis	30-72	11-73	11-73
At time of survey	54.2±11.6	57.0±13.9	56.0±13.0
Female sex	10 (66.7)	16 (64.0)	26 (65.0)
Race/ethnicity			
Asian/Pacific Islanders, non-Hispanic	1 (6.7)	0 (0.0)	1 (2.5)
Black, non-Hispanic	2 (13.3)	0 (0.0)	2 (5.0)
Hispanic	9 (60.0)	0 (0.0)	9 (22.5)
White, non-Hispanic	0 (0.0)	25 (100.0)	25 (62.5)
Others/unknown	3 (20.0)	0 (0.0)	3 (7.5)
Years of health plan enrollment	17.4±13.8	18.5±12.4	18.1±12.8
Currently employed	9 (60.0)	13 (52.0)	22 (55.0)
Charlson Comorbidity Index	0.6±1.1	0.9±1.4	0.8±1.3

POPULATION

This sample of ISM patients is the most racially diverse studied to date; other than race/ethnicity, the two cohorts were similar in other demographic characteristics

Results - Continued

- A lower proportion of Non-White patients (13%) had a long history (≥ 10 years) of diagnosed ISM compared to White patients (32%) **(Table 2)**
- A higher percentage of Non-white patients (47%) were diagnosed 2 or more years following symptom onset than White patients (36%) **(Table 2)**
- A greater proportion of Non-white patients (67%) compared to White patients (40%) reported the journey to be 'moderately or extremely difficult', potentially related to the greater number of physician visits necessary to receive an ISM diagnosis **(Table 2)**
- Less than half (47%) of Non-white patients reported taking medications for ISM compared to 72% of White patients
- A greater proportion of Non-white ISM patients were diagnosed by, and currently receive care from, Hematology or Allergy/Immunology specialists; White patients were more likely to receive care from a wider variety of clinicians

Table 2. Patient-Reported ISM Diagnosis Journey

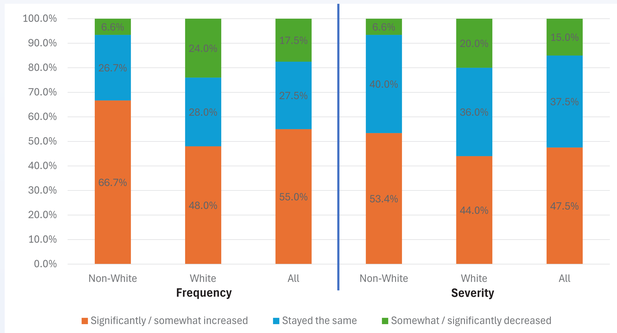
	Race/Ethnicity Group		
	Non-White (N = 15)	White (N = 25)	Total (N = 40)
Time to diagnosis (years)			
<2 years	8 (53.3)	16 (64.0)	24 (60.0)
≥2 years	7 (46.7)	9 (36.0)	16 (40.0)
Visits for symptoms to diagnosis			
1 to 5 visits	7 (46.7)	14 (56.0)	21 (52.5)
6 or more visits	8 (53.3)	11 (44.0)	19 (47.5)
Difficulty to ISM diagnosis			
Easy	3 (20.0)	8 (32.0)	11 (27.5)
Neither easy nor difficult	2 (13.3)	7 (28.0)	9 (22.5)
Moderately or extremely difficult	10 (66.7)	10 (40.0)	20 (50.0)
Duration since diagnosis (years)			
<1	2 (13.3)	3 (12.0)	5 (12.5)
1 to 4	7 (46.7)	6 (24.0)	13 (32.5)
5 to 9	4 (26.7)	8 (32.0)	12 (30.0)
≥10	2 (13.3)	8 (32.0)	10 (25.0)

- Multi-symptom prevalence increase since diagnosis for ISM patients, with skin symptoms as the most prevalent at diagnosis and currently; however, symptomatology at initial presentation may vary by race/ethnicity **(Table 3)**
- Both at diagnosis and time of survey administration, Non-white ISM patients appeared more likely to experience neurocognitive, gastrointestinal, and pain symptoms compared to White patients **(Table 3)**
- After diagnosis, more Non-white patients reported their ISM symptoms had 'somewhat or significantly increased' in frequency and severity (64% and 53%, respectively) compared to White patients (48% and 44%, respectively) **(Figure 1)**
- ISM patients reported worse physical and mental health compared to population norms (Physical Component Score (PCS) = 50.1; Mental Component Score (MCS) = 50.0). This was true for both white (PCS=47.3; MCS=47.6) and Non-white (PCS=45.8; MCS=47.7) patients

Table 3. ISM Symptoms at Diagnosis & Time of Survey (Current)

	Race/Ethnicity Group					
	Non-White (N = 15)		White (N = 25)		Total (N = 40)	
Symptoms present	**At Diagnosis**	**Current**	**At Diagnosis**	**Current**	**At Diagnosis**	**Current**
Skin	13 (86.7)	14 (93.3)	22 (88.0)	23 (92.0)	35 (87.5)	37 (92.5)
GI	8 (53.3)	9 (60.0)	10 (40.0)	13 (52.0)	18 (45.0)	22 (55.0)
Fatigue	8 (53.3)	9 (60.0)	12 (48.0)	13 (52.0)	20 (50.0)	22 (55.0)
Neurological	10 (66.7)	10 (66.7)	14 (56.0)	16 (64.0)	24 (60.0)	26 (65.0)
Pain (muscle, joint, bone)	8 (53.3)	9 (60.0)	9 (36.0)	15 (60.0)	17 (42.5)	24 (60.0)
Respiratory	1 (6.7)	1 (6.7)	2 (8.0)	1 (4.0)	3 (7.5)	2 (5.0)
Bone (osteoporosis/osteopenia)	3 (20.0)	5 (33.3)	7 (28.0)	12 (48.0)	10 (25.0)	17 (42.5)
Anaphylaxis (or severe allergic reaction)	4 (26.7)	3 (20.0)	7 (28.0)	8 (32.0)	11 (27.5)	11 (27.5)

Figure 1. Symptom Change Since Diagnosis



SYMPTOM CHANGE

More Non-white patients reported more frequent and severe ISM symptoms following diagnosis than White patients

Conclusions

- The survey explores racial disparities in the patient burden of ISM
- The journey to ISM diagnosis was longer, more difficult, and associated with more healthcare visits among Non-white patients
- Non-white ISM patients reported more frequent and more severe symptoms following diagnosis
- Symptom presentation and profile over time differed between the two groups, suggesting that disease burden may vary by race and/or ethnicity
- Given the important differences observed in this cohort, research with larger sample sizes in similarly diverse populations is warranted

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