Economic Burden of Macrophage Activation Syndrome (MAS) in Patients With Still's Disease (Systemic Juvenile Idiopathic Arthritis [sJIA] and Adult-onset Still's Disease [AOSD]): Analysis of a US National Administrative Claims Database

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CONCLUSIONS

- **Patients with macrophage activation syndrome (MAS) associated with Still's disease incurred significantly higher healthcare resource utilization (HCRU) and related costs compared with patients with Still's disease without MAS**
- **Novel MAS-targeted treatments may alleviate the overall disease and economic burden on patients and the healthcare system**

BACKGROUND

- MAS is a rare, potentially-fatal, systemic hyperinflammatory syndrome that occurs as a complication of rheumatologic disease, particularly Still's disease (systemic juvenile idiopathic arthritis [sJIA] and adult-onset Still's disease [AOSD])[1,2]
- Untreated or inadequately treated MAS may rapidly progress to multiorgan failure and death[3]
- There is only 1 recently approved therapy for MAS in Still's disease, emapalumab; however; the economic burden (HCRU and costs) of MAS in patients with Still's disease in the United States (US) is unknown[4]

OBJECTIVE

To assess HCRU and costs associated with MAS in patients with Still's disease using a US national administrative claims database

METHODS

- In this retrospective analysis, the IQVIA PharMetrics Plus administrative claims database from 2019 to 2024 was used to identify:
  - Patients with Still's disease (International Classification of Diseases [ICD]-10: M082), including AOSD (ICD-10: M061) and sJIA (ICD-10: M08) with ≥6 months of continuous enrollment prior to Still's disease diagnosis (index date) and ≥12 months of continuous enrollment post-index without a MAS diagnosis (ICD-10:D76.1)
  - Patients with Still's disease with ≥6 months of continuous enrollment prior to earliest MAS diagnosis (index date) observed in the study period and ≥12 months of continuous enrollment post-MAS diagnosis
- A propensity score (PS) was used to match patients (1:1) with Still's disease with and without MAS. Baseline variables used for PS matching were age at Still's disease diagnosis, sex, race/ethnicity, underlying Still's disease diagnosis (AOSD or sJIA), and Elixhauser Comorbidity Index (ECI)
- Odds ratios (ORs) were evaluated for binary variables (≥1 visit) using logistic regression
- Generalized linear models were used to estimate the impact of MAS on HCRU (Poisson distribution) and costs (gamma distribution)
- Mean differences in HCRU and costs between the 2 groups were estimated using unadjusted models and models that adjusted for age at Still's disease diagnosis, underlying Still's disease diagnosis, sex, and ECI during the baseline period

RESULTS

- After PS matching, 45 patients with Still's disease and MAS and 45 patients with Still's disease only were included in the analysis
- In both cohorts, the mean age at diagnosis of Still's disease was 26.8 years, more than one-half of the patients were female, and more (>60%) patients had AOSD than sJIA **(Table 1)**
- MAS was diagnosed at a mean (standard deviation [SD]) of 4.6 (10.1) years after Still's disease diagnosis
- The mean (SD) ECI was higher in patients with Still's disease and MAS than in patients with Still's disease without MAS **(Table 1)**
- In both cohorts, >40% of the patients had 1 to 2 comorbidities. The most common comorbidities were rheumatoid arthritis and cardiac arrythmia **(Table 1)**

Table 1. Patient demographics and clinical characteristics at Still's disease diagnosis

	Still's disease with MAS (n = 45)	Still's disease without MAS (n = 45)
Age at diagnosis of Still's disease, mean (SD), years	26.8 (16.0)	26.8 (15.1)
Sex, n (%)		
Female	27 (60.0)	25 (55.6)
Still's disease diagnosis		
Adult-onset Still's disease, n (%)	30 (66.7)	31 (68.9)
Systemic juvenile idiopathic arthritis, n (%)	15 (33.3)	14 (31.1)
Elixhauser Comorbidity Index, mean (SD)	2.1 (2.8)	1.6 (1.5)
Elixhauser comorbidities, n (%)		
Rheumatoid arthritis	19 (42.2)	22 (48.9)
Fluid and electrolyte disorder	12 (26.7)	3 (6.7)
Cardiac arrhythmia	10 (22.2)	6 (13.3)
Depression	9 (7.3)	17 (13.7)
Anemia deficiency	3 (6.7)	7 (15.6)
Type of insurance, n (%)		
Commercial insurance	16 (35.6)	21 (46.7)
Self-insured	28 (62.2)	22 (48.9)

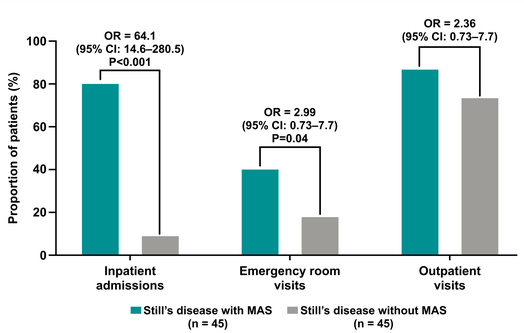
MAS, macrophage activation syndrome; SD, standard deviation.

- Most patients in both cohorts were self-insured or had commercial insurance and were covered by a Preferred Provider Organization

All-cause HCRU

- In the 12-month post-index period, patients with Still's disease and MAS had significantly more HCRU, including inpatient (IP) admissions (OR: 64.1; P<0.001) and/or emergency room (ER) visits (OR: 2.99; P=0.04) than patients without MAS **(Figure 1)**

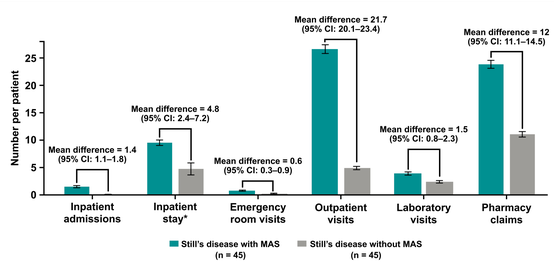
Figure 1. All-cause HCRU (proportion of patients) during the post-index period



CI, confidence interval; HCRU, healthcare resource utilization; MAS, macrophage activation syndrome; OR, odds ratio.

- Patients with Still's disease and MAS had significantly more frequent IP admissions (P<0.001), ER visits (P<0.001), outpatient (OP) visits (P<0.001), and longer IP stays (P<0.001) than patients with Still's disease without MAS **(Figure 2)**
- These results remained robust and did not change when adjusted for age at Still's disease diagnosis, underlying Still's disease diagnosis (AOSD, or sJIA), sex, and ECI

Figure 2. All-cause HCRU during the post-index period

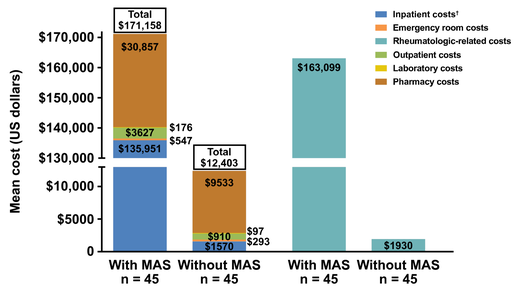


*Limited to patients with ≥1 inpatient admission. P-values <0.001 for comparisons across all types of visits.
CI, confidence interval; HCRU, healthcare resource utilization; MAS, macrophage activation syndrome.

All-cause costs and rheumatologic-disease associated costs

- Patients with Still's disease and MAS incurred significantly higher total all-cause ($318,828 vs $19,233; P=0.008) and rheumatologic-related ($163,099 vs $1930; P=0.02) healthcare costs than patients without MAS **(Figure 3)**
- These results remained robust and did not change when adjusted for age at Still's disease diagnosis, underlying Still's disease diagnosis (AOSD, or sJIA), sex, and ECI

Figure 3. All-cause healthcare costs* (US dollars) in the post-index period



*Adjusted to 2024 US dollars using the medical component of the Consumer Price Index.
†Inpatient costs limited to patients with ≥1 inpatient admission. MAS, macrophage activation syndrome.

All-cause HCRU and costs 6 months pre- and post-MAS diagnosis

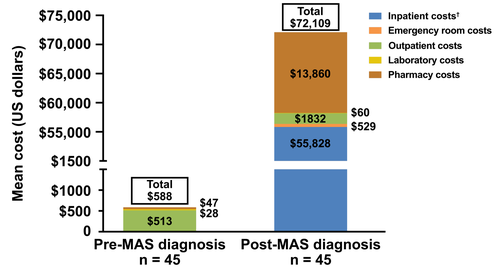
- Patients with Still's disease had significantly higher HCRU **(Table 2)** and all-cause healthcare costs after progressing to MAS than when they had Still's disease only ($71,919 vs $5957; **Figure 4)**

Table 2. All-cause HCRU 6 months pre- and post-MAS diagnosis

	Pre-MAS (n = 45)	Post-MAS (n = 45)	P-value
Inpatient admissions, n			
Median (Q1, Q3)	0 (0, 1)	1 (1, 2)	<0.001
Inpatient length of stay,* (days)			
Median (Q1, Q3)	7 (5.5, 11)	9 (5.5, 12)	0.5
Emergency room visits, n			
Median (Q1, Q3)	0 (0, 1)	0 (0, 1)	1.0
Outpatient visits, n			
Median (Q1, Q3)	4 (1, 8)	11 (4, 26)	<0.001
Laboratory visits, n			
Median (Q1, Q3)	0 (0, 2)	1 (0, 2)	0.007
Pharmacy claims, n			
Median (Q1, Q3)	0 (0, 6)	5 (0, 19)	<0.001

*Limited to patients with ≥1 inpatient admission.
HCRU, healthcare resource utilization; MAS, macrophage activation syndrome; Q1, first quartile; Q3, third quartile.

Figure 4. All-cause healthcare costs* (US dollars) 6 months pre- and post-MAS diagnosis



*Adjusted to 2024 US dollars using the medical component of the Consumer Price Index.
†Inpatient costs limited to patients with ≥1 inpatient admission. MAS, macrophage activation syndrome.

LIMITATIONS

- As with other retrospective studies, there is a risk of missing or incomplete information
- PS matching is limited to the baseline variables for which data are available

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CONFLICTS OF INTEREST

Dr Marrone, Dr McPherson, and Dr Oladapo are employees of Sobi, Inc.

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