Patients' perspective on the burden of Hypereosinophilic Syndrome

Poster No. P247

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Introduction

HES is a group of rare hematologic disorders characterized by hypereosinophilia (typically >1500 cells/µL on two or more occasions), eosinophil-driven organ damage and dysfunction, and exclusion of secondary causes of eosinophilia.¹⁻⁵

The identification and diagnosis of HES is challenging owing to the rarity of the disease, the varied clinical presentation, the lack of recognition and overlapping characteristics with other eosinophilic conditions such as EGPA.⁴⁻⁷

The prevalence of HES is relatively low (0.32 to 6.3 cases per 100,000 people in the US), and the patient journey remains poorly understood.8 A better understanding of the patient burden associated with HES may facilitate understanding of unmet patient needs in HES and thereby direct improvements in diagnosis and management.

This study aimed to describe the experience of patients with HES, from the patients' own perspective from initial symptoms, through diagnosis, to QoL burdens and treatment experiences and was performed in conjunction with the key patient advocacy group, APFED.

Most comm conditions Methods Anxiety Chronic Gastrointe Study design (GSK ID: 214158) COPD, e Chronic s Vasculitis/ U U U U U **Real world Cross-sectional Online survey Fielding period** Patient eligibility criteria* ≥18 years of age Feb 2022 to 2022 Self-reported diagnosis of HES 65% of oatients too longer tha Identification of Primary target 6 months fo survey respondents rvey population diagnosi Email and social media reach-outs via APFED Patients with diagnosed HES and their (patient advocacy group for patients with HES) caregivers as identified through: APFED's email listserv 200 Patient recruitment social media platforms Through APFED's opt-in patient database and a atients recruited patient registry platform that APFED partners from the US: with[†] (joined at-will by patients after learning about the network)[‡] N=54 Exploratory analysis endpoints L. 299 Diagnostic Patient Symptom Treatment history Disease impact history burden on QoL§ demographics and burden and disease characteristics

Caregivers of eligible patients could complete the survey on the patient's behalf; †Eosinophil Connect Patient Insights Network hosted on the Invitae platform; [‡]Through their providers, internet searches, support groups/platforms, social media, news outlets and word of mouth; §The impact of HES on patient's QoL was described using the following variables: Impact on activities of daily living (including intensity and duration) includes daily impact, work/school, recreation, other activities, social/relationships, outings, other; and most impactful QoL activities.

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Results

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Table 1. The majority of patients had idiopathic HES, and asthma was the most common comorbid or associated condition						
		Overall (N=54)			Overall (N=54)	
	Age, mean (SD), years	43.6 (14.0)		Eosinophilic complications, n (%) Respiratory [†] Gastrointestinal [‡] Vascular [§]	14 (26)	
	Male, n (%)	31 (57.4)			31 (57) 7 (13)	
	HES subtype, n (%) Idiopathic Myeloid variant Lymphocytic variant Other/unsure	32 (59) 15 (28) 3 (6) 5 (9)		Five most common treatments taken prior to diagnosis, n (%) Steroids Monoclonal antibody injectable medications Non-prescription management Hydroxyurea Vincristine	28 (52) 15 (28) 12 (22)	
	Family history of eosinophilic disorders, n (%)	18 (33)			9 (17) 8 (15)	
%	Most common comorbid or associated conditions, n (%) Asthma Anxiety Chronic skin disease Gastrointestinal disorders* COPD, emphysema, chronic bronchitis Chronic sinusitis Vasculitis/EGPA	29 (54) 14 (26) 13 (24) 12 (22) 8 (15) 7 (13) 7 (13)		HES-related HCRU in the past 12 months, n (%) Primary care provider Urgent care Emergency room Hospital admission Allergist/immunologist Pulmonologist Other specialist**	27 (50) 3 (6) 9 (17) 27 (50) 26 (48) 16 (30) 15 (28)	

*Includes esophagitis, gastritis, colitis, irritable bowel disease; †Includes COPD, current/former smoker, eosinophilic asthma and eosinophilic pneumonia; ‡Includes eosinophilic gastritis/gastroenteritis, eosinophilic esophagitis, eosinophilic duodenitis and eosinophilic colitis; §Includes EGPA; Includes over-the-counter medications: Tylenol and Flonase nasal spray; **Includes gastroenterologists, ophthalmologists, hematologists, oncologists, and NIH.





*Includes cardiologist, gastroenterologist and neurologist; †Includes spinal tap, endoscopy, colonoscopy, psychological tests, pulmonary function tests

Abbreviations

APFED, American Partnership for Eosinophilic Disorders, COPD, chronic obstructive pulmonary disease; CT, computerized tomography; EGPA, eosinophilic granulomatosis with polyangiitis; HCRU, healthcare resource use; HES, hypereosinophilic syndrome; MRI, magnetic resonance imaging; NIH, National Institute of Health; QoL, quality of life; SD, standard deviation; US, United States.

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Figure 2. Patients with HES experienced a broad range of clinical symptoms* Conclusions Circulatory symptoms, % 🗾 Integumentary symptoms, % • This study, using data collected through an exploratory patient survey, demonstrates the heterogeneous clinical Heart palpitations/irregular heartbeat presentation of HES and highlights the multifactorial burden associated with this disease. Chest pain Itching - Patients reported that the diagnosis pathway was complex, with several months to years often passing Redness of the skin between the first symptoms and diagnosis, and over a third found the diagnostic process very difficult. This Patches of scaly skin likely reflects patients visiting multiple healthcare providers and having extensive diagnostic testing. - Idiopathic HES was the most common variant. The rate of myeloid HES was higher and lymphocytic HES **F** Digestive symptoms, % lower than previously documented, where incidences were reported between 10% and 20% for both Respiratory symptoms, % Nausea variants.5,9-11 Diarrhea Wheezing The range of symptoms experienced by patients was broad, with the disease affecting multiple organ Difficulty swallowing Dry cough systems, and the management of which took time away from day-to-day activities; many reported adverse Shortness of breath Pain in the abdomer Lack of appetite Chest tightness impacts on their QoL. Stuffv nose Monoclonal antibody therapy was the most commonly used current HES treatment, and was associated with symptomatic control, and linked closely with improved QoL for these patients. Nervous system symptoms, % - Most patients were satisfied with their steroid treatment, and half felt the steroids helped improve their QoL Muscular symptoms, 9 but the long-term adverse effects associated with their use were not explored.⁴ Muscle pair Headache General symptoms, % • These data highlight the key unmet needs of this population and offer a basis upon which diagnosis, care, and Dizziness Joint pain 00 Fatigue Muscle spasms 33 Numbness in hands/feet outcomes may be improved. General discomfort Joint swelling Difficulty concentrating Fever Weakness in arms/legs Weakness Memory problems Weight loss Figure 4. The most common QoL impacts adversely affecting patients included ability to exercise, work quality, ability to participate in social activities and anxiety/worry, and the overall HES burden was high *Where possible, the five most common symptoms in each category are presented. Symptoms may have been reported in more than one category, for example, shortness of breath and dizziness were also captured under circulatory symptoms (22%). QoL impacts* 100 Figure 3. Patients reported a substantial treatment burden associated with HES, but satisfaction with monoclonal antibody ijectable medications was high Overall (N=54) I am satisfied with how this treatment controlled/controls my Time from HES diagnosis to HES symptoms (somewhat agree + strongly agree)[‡], n (%) treatment initiation, n (%) Monoclonal antibody injectable medications* (N=25) 17 (68) 30 (56) <3 months Hydroxyurea (N=28) 19 (68) 12 (22) 3-6 months Steroids (N=38) 26 (68) 5 (9) 3 (6) 3 (6) 1 (2) 6–12 months 4 (31) Non-prescription management[†] (N=13) 1–3 years Cyclosporine (N=11) 3 (27) 3–5 years ≥5 years Five most common current HES I feel like this treatment helped/helps improve my QoL treatments, n (%) (somewhat agree + strongly agree)[‡], n (%) 19 (35) Monoclonal antibody injectable Monoclonal antibody injectable medications* (N=25) 18 (72) medications' 15 (54) Hydroxyurea (N=28) Other activities Daily activities Work/school activities Social life/relationship activities 13 (24) Hydroxyurea Steroids (N=40) 20 (50) 12 (22) Steroids 3 (23) Non-prescription management[†] (N=13) **Overall HES burde** 9 (17) Non-prescription management[†] Cyclosporine (N=11) 6 (55) 8 (15) Cyclosporine Strongly disagree Treatment burden: I feel that taking care of my HES takes time In general, this treatment is/was overall easy to manage Somewhat disagree away from other activities, n (%) Due to my (somewhat agree + strongly agree)[‡], n (%) HES, I feel I feel my HES I feel different Strongly disagree Monoclonal antibody injectable medications* (N=25) 19 (76) unable to do 3 (6) from others/ symptoms are Somewhat disagree Hydroxyurea (N=28) 22 (79) Somewhat agree certain things 14 (26) misunderstood burdensome Neither agree nor disagree Steroids (N=40) 26 (65) for/with 20 (37) Somewhat agree Strongly agree 7 (54) Non-prescription management[†] (N=13) my family 16 (30) Strongly agree 7 (64) Cyclosporine (N=11) 14 (26) N/A or prefer not to answer 'Includes etoposide 50 mg tablet; †Includes over-the-counter medications: Tylenol and Flonase nasal spray; ‡Data shown for the five most common current HES treatments. *The seven most common QoL impacts in each category are shown; *Percentage of patients in each category depicted.

• On behalf of all authors, and with their permission, an audio recording of this poster was

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