Request for ICD-10-CM Codes for Eosinophil-Associated Diseases

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Disclosures for Dr. Bochner

- I currently serve as a consultant for Genentech.
- I receive research funding from the NIH and from Acerta Pharma.
- I am the President of the International Eosinophil Society, for which I receive no remuneration.
- I receive publishing royalties from Elsevier and Wolters Kluwer/UpToDate.
- I am a co-founder, own stock in, and serve on the scientific advisory board of Allakos Inc., a biopharmaceutical company developing antibodies to Siglec-8 for therapeutic use.
- I am a co-inventor on Siglec-8-related patents owned by the Johns Hopkins University, and am entitled to a share of future royalties received by the University on eventual sales of products protected by those patents.
- The terms of these arrangements are being managed by the Johns Hopkins University and Northwestern University in accordance with their conflict of interest policies.

The International Eosinophil Society, Inc. (IES) and American Partnership for Eosinophilic Disorders (APFED) wrote this proposal.

IES is an organization of scientists and clinicians interested in the eosinophil, a blood cell strongly associated with many diseases. The society sponsors biennial meetings (next one in July 2019) to review new information about the eosinophil and its roles in health and disease.

APFED is a 501c3 nonprofit organization founded in December 2001. APFED's mission is to passionately embrace, support, and improve the lives of patients and families affected by eosinophil-associated diseases through education and awareness, research, support, and advocacy.

International Eosinophil Society
IES

Apfed
American Partnership
for Eosinophilic Disorders

ICD-10-CM: Request for 8 New Eosinophil-Associated Diseases (EADs) Codes and 4 Amended Codes

APFED requested and the CDC approved in 2007 **four ICD-9 codes for eosinophil gastrointestinal diseases** (EGIDs). These were converted to **three ICD-10-CM codes**.

Other EADs generally use two generic "catch-all" codes, **J82** (pulmonary eosinophilia, not elsewhere classified) and **D72.1** (eosinophilia), either in isolation or in combination with other non-specific codes.

Four "amendments" to existing codes, including a name change [Churg Strauss syndrome to EPGA], a separation of two codes that were approved as unique codes in ICD-9 [EG/EGE], and add exclusions to an existing code [EC].

Eight new EAD codes requested**.**

Without ICD-10-CM Codes for EADs…?

- Extended time to diagnosis
- Unmet patient needs in management and treatment options in these EADs because each is a different disorder with unique pathophysiology
- Healthcare resources misallocated given current prevalence and inability to collect patient care data and estimate “real” cost of care
- Unable to develop treatments or provide access to currently available approved treatments
- Missed opportunities to identify potential clinical study recruits

Requesting ICD-10-CM Codes for these EADs

Pulmonary Eosinophil Diseases

- ❑ Acute Eosinophilic Pneumonia
- ❑ Chronic Eosinophilic Pneumonia
- ❑ Eosinophilic Asthma

Hypereosinophilic Syndromes

- ❑ Myeloid Hypereosinophilic Syndrome (MHES)
- ❑ Lymphocytic Variant Hypereosinophilic Syndrome (LHES)
- ❑ Idiopathic Hypereosinophilic Syndrome (IHES)
- ❑ Episodic Angioedema with Eosinophilia (EAE), also called Gleich's Syndrome

Other

- ❑ Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS)
- ❑ Eosinophilic Granulomatosis with Polyangiitis (EGPA), formerly called Churg-Strauss Syndrome*

EGIDs

- ❑ Eosinophilic Gastritis (EG)*
- ❑ Eosinophilic Gastroenteritis (EGE)*
- ❑ Eosinophilic Colitis (EC)*

*REQUESTING AMENDMENTS OR NAME CHANGE ONLY

Acute versus Chronic Eosinophilic Pneumonia

- **Acute Eosinophilic Pneumonia (AEP)** was first described as a distinct entity in medical literature in 1989.
- AEP is characterized by rapidly progressive respiratory failure with high levels of lung eosinophils (>25%).
- Following diagnosis and corticosteroid treatment, prognosis is excellent.
- Cause is unknown but smoking and environment may be triggers (e.g., 9/11 rescue workers with dust exposure).
- Currently Using Code J18. (for various pneumonias) and/or J82 (pulmonary eosinophilia, not elsewhere classified)

- **Chronic Eosinophilic Pneumonia (CEP)** was first described in 1969 and is a distinct entity.
- CEP is characterized by progressive shortness of breath and increase in eosinophils in the lungs and bloodstream; abnormalities on chest imaging in the periphery of the lungs, but unlike AEP does not progress to acute respiratory failure.
- Relapse over many years is common even with treatment. Can progress to severe asthma or EGPA.
- Cause is unknown; more common in women and adults.
- Currently Using Code J18. (for various pneumonias) and/or J82 (pulmonary eosinophilia, not elsewhere classified)

Eosinophilic Asthma (currently coded as J45.* with J82 rarely added)

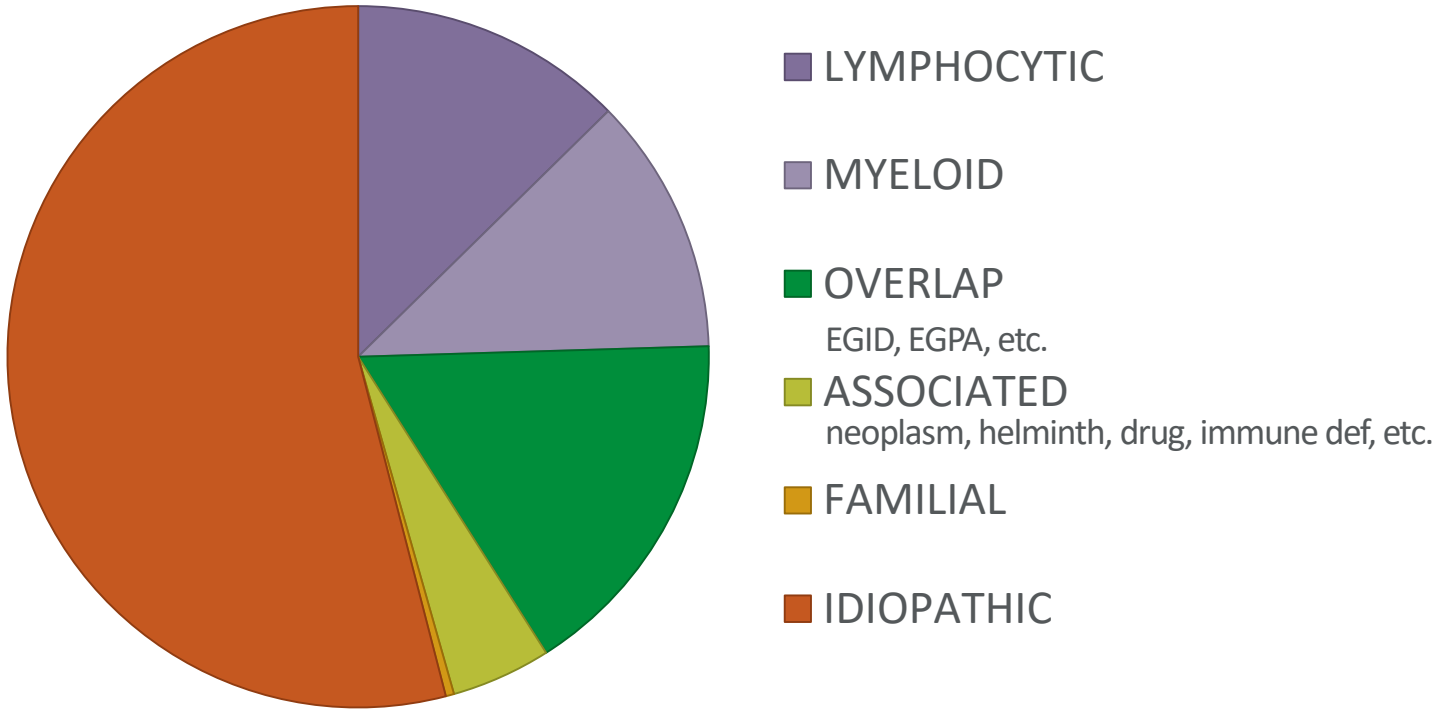
- Cited in research since 1889. Subset of asthma that is characterized by the presence of eosinophils in the circulation and in the airways.
- Is considered a leading cause of severe asthma, affecting 50 to 60 percent (approx 2 million) of those with severe asthma.
- Often associated with chronic sinusitis and nasal polyposis.
- Is more prominent in adults and equally affects men and women (different from other forms of adult asthma).
- Frequent exacerbations and detrimental effect on QOL. Does not respond well to inhaled corticosteroid treatments, even at high doses.
- Three new FDA approved biologic therapies that specifically target eosinophils (by blocking IL-5 or its receptor) reduce asthma exacerbations and are oral steroid sparing.

Hypereosinophilic Syndromes (HES): *MHES, LHES, IHES, EAE*

- HES are rare disorders, with an estimated prevalence of 1.5-3 per 100,000.
- HES subtype has important implications both for natural history, management, and prognosis.
- HES are characterized by an elevated blood eosinophil count and eosinophil-mediated end-organ damage (most commonly cutaneous, pulmonary, gastrointestinal, cardiac, and nervous system tissues).
- Over the past 20 years, considerable progress has been made in understanding the pathogenesis and improving treatment of HES subtypes, but each is different.
- The ongoing clinical development of kinase inhibitors (e.g. imatinib), biologics (e.g. mepolizumab, reslizumab, benralizumab, dupilumab) and other therapies in HES will continue to validate a precision medicine approach, with each drug having its greatest clinical benefit limited to specific HES subpopulations.

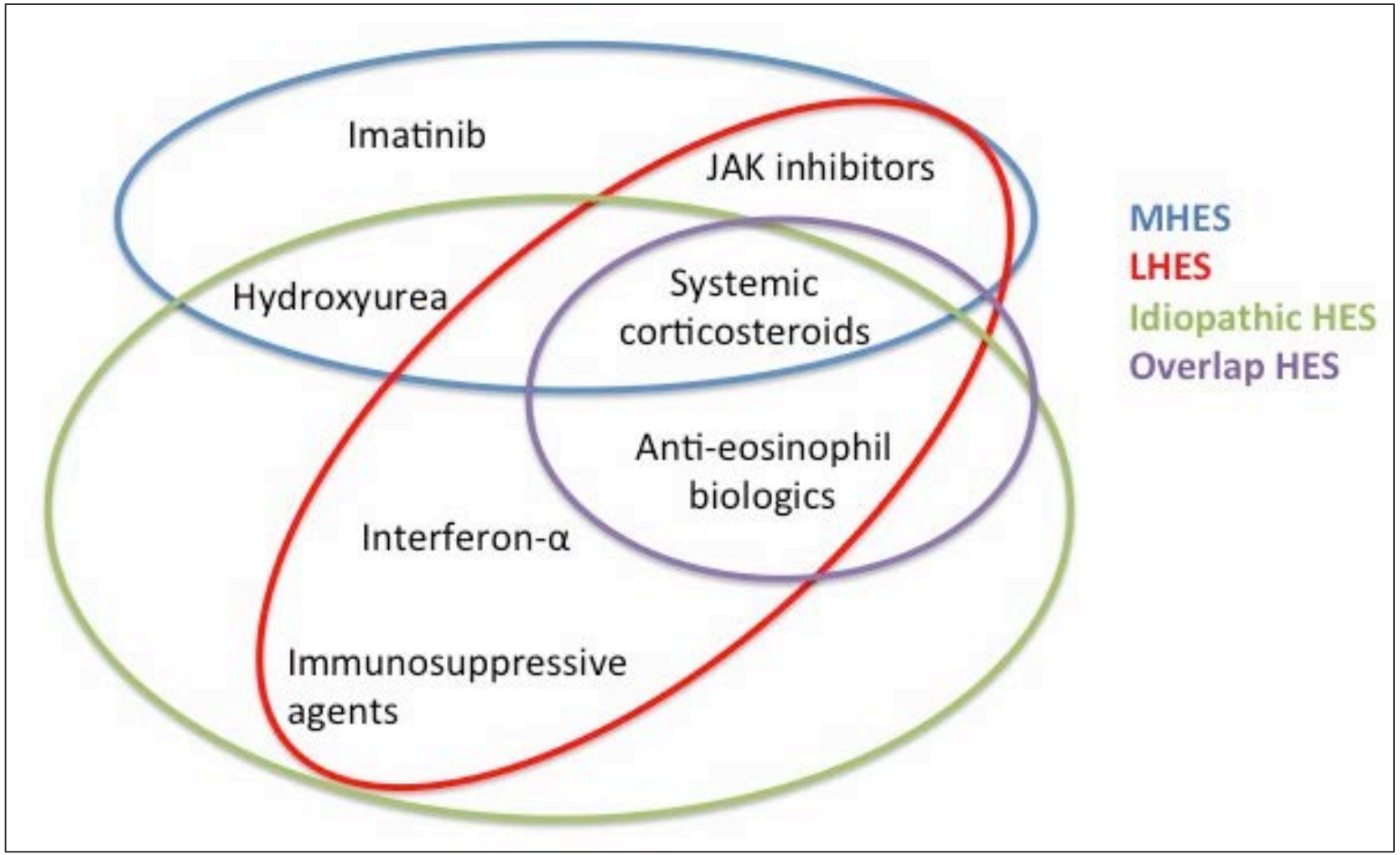
Klion, Hematology Am Soc Hematol Educ Program 2018:326-31, 2018

Frequency of clinical subtypes of HES



Klion, Blood 126:1069-77, 2015

Treatments for Hypereosinophilic Syndromes



Khoury and Bochner, J Allergy Clin Immunol Pract 6:1446-53, 2018

Hypereosinophilic Syndromes (HES)

Myeloid Hypereosinophilic Syndrome (MHES)

- Currently Using Code **D72.1 (“eosinophilia”)** but this is really eosinophilic leukemia
- Approximately 50% of MHES (5-8% of all HES) is caused by a FIP1L1-PDGFRA fusion gene mutation on chromosome 4, and is **cured** with tyrosine kinase inhibitors like imatinib.
- An additional population of approximately 10-20% of MHES is associated with other fusion genes or mutations involving PDGFRA, PDGFRB, FGFR1, or JAK2.

Lymphocytic Variant Hypereosinophilic Syndrome (LHES)

- Currently Using Code **D72.1 (“eosinophilia”)**
- LHES accounts for approximately 15% of HES and is defined by the presence of clonal and/or aberrant T lymphocytes that produce Th2 cytokines, such as interleukin-5, that drive eosinophilia.
- Rash with severe itching is the most common presentation.
- LHES may progress into lymphoma in 5-25% of patients.

Hypereosinophilic Syndromes (HES)

Idiopathic Hypereosinophilic Syndrome (IHES)

- Currently Using Code **D72.1 (“eosinophilia”)** but this may also sometimes be eosinophilic leukemia
- IHES accounts for approximately 70% of HES.
- Cause of IHES is unknown, and may affect any organ including the heart.
- Corticosteroids are the first line treatment.
- Currently no approved drugs indicated for treatment.

Episodic Angioedema with Eosinophilia (EAE), Gleich’s Syndrome

- Currently Using Code **D72.1 (“eosinophilia”)**
- First described in 1984 as a cyclic disorder characterized by recurrent episodes of fever, swelling, weight gain and eosinophilia recurring every 4-6 weeks.
- EAE makes up <1% of patients with HES.
- EAE has traditionally been considered a variant of HES, and more recently LHES.
- Disease onset may occur at any age but is more common in adulthood.

Other Eosinophil-Associated Diseases

Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS)

- Currently Using Code: **D72.1** (“eosinophilia”) + **T88.7** (“unspecified adverse effect of a drug or medicament”)
- ICD-11 Code EH65

Eosinophilic Granulomatosis with Polyangiitis (EGPA) previously known as Polyarteritis with Lung Involvement [Churg-Strauss Syndrome]

- Name Change Only
- Currently Using Code: **M30.1**
- ICD-11 Code 4A44.A2

Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS)

- Idiosyncratic reaction to the use of certain drugs.
- Pathophysiology unknown, but is associated with immunological or genetic factors.
- Incidence is 0.4 cases per 1,000,000 in the general population.
- Rash typically starts 2-8 weeks AFTER starting a drug, like an anti-epileptic or antibiotic.
- In addition to the rash, fevers, eosinophilia and systemic manifestations occur (e.g., lymphadenopathy; viral reactivation; kidney, liver, lung involvement) and take months to resolve.
- Unlike common drug rashes, DRESS is quite serious and death can occur.
- Therapeutic management includes stopping the suspected drug and applying empiric treatment, including symptomatic management, life support and use of corticosteroids.
- The lack of an ICD code has severely hampered the ability to assess pharmacogenetics, detect new culprit agents, and assess important immunopathogenic questions regarding etiology, impact of genetic background, and potential for cross-reactive medications.

Eosinophilic Granulomatosis with Polyangiitis (EGPA)

Previously Known as Churg-Strauss Syndrome (Polyarteritis with lung involvement)

- Requesting name change only- Eosinophilic Granulomatosis with Polyangiitis (EGPA) as the official name of what had previously referred to as Churg-Strauss Syndrome.
- Is a rare autoimmune disorder that may affect multiple organ systems, especially the lungs in the form of asthma, causing damage.
- Characterized by abnormal presence of high blood and tissue eosinophils, inflammation of blood vessels (vasculitis), and development of inflammatory nodular lesions called granulomas (granulomatosis).
- Without treatment, serious organ damage can occur and the disease may be fatal.
- The cause of EGPA is unknown.

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Eosinophil Gastrointestinal Diseases (EGIDs)

Code Amendments requested

Eosinophilic Gastritis (EG) and Eosinophilic Gastroenteritis (EGE)

- In 2008, the CDC CM Committee approved ICD-9 Codes for Eosinophilic Gastroenteritis (558.41) and Eosinophilic Gastritis (535.7). In the transition to ICD-10, the codes for these distinct diseases were combined.
- Currently using same code for both EG and EGE: **K52.81**
- ICD-11 has two distinct codes for EG: DA94.21 and EGE DA42.2

Eosinophil Gastrointestinal Diseases (EGIDs)

Code Amendments requested

Eosinophilic Colitis

- Existing Code: **K52.82**
- Request to **delete inclusions for food protein induced enterocolitis syndrome (FPIES), allergic proctocolitis and milk-protein colitis**.
- In 2016, FPIES was approved for new ICD-10 CM code **K52.21,** so we are requesting an **exclusion for FPIES be added to K52.82**.
- Eosinophilic colitis diagnoses requires colonoscopy showing elevated eosinophils. Allergic proctocolitis and milk-protein colitis are not diagnosed with colonoscopy, occur in newborns, and are usually self-resolving diseases.

Summary: Request for ICD-10-CM Codes for Eosinophil-Associated Diseases

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Eight new EAD codes requested.

IES and APFED appreciate the opportunity to present this proposal to the CDC CMS Committee

Apfed

American Partnership
for Eosinophilic Disorders