New Allergic and Hypersensitivity Conditions Section in the International Classification of Diseases–11

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INTRODUCTION

Allergy and hypersensitivity, originally perceived as rare and secondary disorders, are one of the fastest growing conditions worldwide, but not adequately tracked in international information systems, such as the International Classification of Diseases (ICD). Having allergic and hypersensitivity conditions classification able to capture conditions in health international information systems in a realistic manner is crucial to the identification of potential problems, and in a wider system, can identify contextually specific service deficiencies and provide the impetus for changes. Since 2013, an international collaboration of Allergy Academies has spent tremendous efforts to have a better and updated classification of allergies in the forthcoming International Classification of Diseases (ICD)-11 version, by providing scientific and technical evidences for the need for changes. The following bilateral discussions with the representatives of the ICD-11 revision, a simplification process was carried out. The new parented “Allergic and hypersensitivity conditions” section has been built under the “Disorders of the Immune System” chapter through the international collaboration of Allergy Academies and upon ICD WHO representatives support. The classification of allergic and hypersensitivity conditions has been updated through the ICD-11 revision and will allow the aggregation of reliable data to perform positive quality-improvements in health care systems worldwide.

Key Words: Allergic disorders; allergy; hypersensitivity; classification; international classification of diseases; world health organization

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provide reliable data, decreasing the visibility of some conditions in detriment to the others, there is a possibility of negative outcomes in health decision-making and management actions, affecting the supply and demand of goods and services in both national and global levels. This also results in poor understanding of their natural history and lack of knowledge of their epidemiology.

MATERIALS AND METHODS

Considering the ongoing ICD-11 revision an unique opportunity to standardize coding definitions not just for anaphylaxis but for all hypersensitivity/allergic disorders, we organized an international collaboration of regional Allergy Academies, first including the European Academy of Allergy and Clinical Immunology (EAACI), the World Allergy Organization (WAO), the American Academy of Allergy Asthma and Immunology (AAAAI) and then the Latin American Society of Allergy, Asthma and Immunology (SLAAI), the Asia Pacific Association of Allergy, Asthma and Clinical Immunology (APAAACI), and the American College of Allergy, Asthma and Immunology (ACAAI). The joint Allergy Academies have been coordinating a strategic action plan and tremendous efforts since 2013 to provide a better classification of these disorders in the new ICD-11. We first conducted an international survey and demonstrated that the ICD is the most frequently used classification system by the allergy community worldwide; however, it was not considered appropriate in clinical practice.

Early bilateral discussions with the representatives of the ICD-11 revision group have been put in place, and comments have been submitted to the ICD-11 beta draft platform. A careful comparison between ICD-10 and ICD-11 beta phase for allergy/hypersensitivity conditions codes allowed the identification of gaps and trade-offs and supported the construction of a classification proposal. The building process of this model was delineated by ICD/WHO rules, updated by the most recent publications and with the aim to be used by allergists, non-allergists and non-physicians. This classification proposal was validated by crowdsourcing allergist leaderships’ community.

The high level complex structure underwent a cross-linking terms process to coordinate aligning the allergic and hypersensitivity conditions classification to the ICD-11 beta draft facilitating the classification proposal acceptance. The proposed model has been presented to the WHO groups in charge of the ICD revision in December 2014. The strategies used and the collaboration from Academies were acknowledged, and the classification proposal has been well accepted. Advised by these groups, we performed a technical process of simplification in an attempt to better fit it to the ICD-11 framework.

WHO asked to simplify the document to facilitate the alignment of the allergic and hypersensitivity conditions classification ICD-11 beta draft facilitating its acceptance. The final simplified version counted by using 215 terms, but kept the same original structure and the philosophy used for its construction.

RESULTS

The main outcome of the process was the offer to include a section addressed to “allergic and hypersensitivity conditions” (Table) into the ICD-11 framework, allowing a big picture of these conditions, previously undernotified or misclassified in global health registries. The tuned version of the allergic and hypersensitivity conditions classification as well as the list of missing terms endorsed by WHO and related Topic Advisory Groups (TAGs) (Pediatric, Dermatology, Rare Diseases, Ophthalmology, Internal Medicine) was the basis of the construction of the new “Allergic and hypersensitivity conditions” section parented under the “Disorders of the Immune System” chapter (Table). Upon WHO guidance, all the proposals have been submitted into the ICD-11 beta draft platform and during this process, with the aim of reaching a harmonized view regarding overlapping conditions, we have been in contact with relevant WHO TAGs and working groups (WG). Once the classification is included into the ICD-11 framework, our aim is to carry on working in collaboration with WHO in order to validate/disseminate the classification by field trial.

DISCUSSION

Having a classification able to capture conditions in health international information systems in a realistic manner is crucial to the identification of potential problems, and in a wider system, can identify contextually specific service deficiencies and provide the impetus for changes. The new allergic and hypersensitivity conditions section into the ICD-11 framework gave a unit for a specialty previously considered with less importance. Since most of the allergic and hypersensitivity conditions have been considered noncommunicable diseases, the WHO has been cosigning initiatives to support changes in the same direction, such as for the asthma under the Global Alliance against Chronic Respiratory Diseases (GARD) or for the nomenclature of allergens under the WHO/International Union of Immunological Societies (IIUIS). These changes will allow us to monitor the balance between health and allergic/hypersensitivity disease worldwide to understand public policies required to support organized high-impact measures and affordable interventions to prevent, promote health by assuring the access to appropriate care, and improve the quality of life of the population as a whole.

The final ICD-11 framework will be presented to the World Health Assembly in the next few years. We are aware that the revision process is not set and that the current structure may be
### Table. The new “Allergic and hypersensitivity conditions” ICD-11 chapter

| **Allergic or hypersensitivity disorders involving the respiratory tract** | **Asthma** | **Allergic Aspergillus rhinosinusitis** |
| --- | --- | --- |
| Allergic and non-allergic rhinitis | Allergic rhinitis | Bird fancier lung |
| Allergic rhinitis | Allergic rhinitis due to allergens | Suberosis |
| Allergic rhinitis due to allergens | Allergic rhinitis due to pollen | Maltworker lung |
| Allergic rhinitis due to pollen | Allergic rhinitis due to house dust mite | Mushroom-worker lung |
| Allergic rhinitis due to house dust mite | Other allergic rhinitis due to allergens | Maple-bark-stripper lung |
| Other allergic rhinitis due to allergens | Other allergic rhinitis | Air-conditioner and humidifier lung |
| Other allergic rhinitis | Non-allergic rhinitis | Cheese washer's lung |
| Non-allergic rhinitis | Gustatory rhinitis | Coffee worker's lung |
| Gustatory rhinitis | Hormonal-induced rhinitis | Fishmeal worker's lung |
| Hormonal-induced rhinitis | Rhinitis related to pregnancy | Grainhandler’s disease or lung |
| Rhinitis related to pregnancy | Rhinitis related to hypothyroidism | Pituitary-snuff-taker's disease |
| Rhinitis related to hypothyroidism | Drug-induced rhinitis | Red-cedar lung or pneumonitis |
| Drug-induced rhinitis | Non-allergic rhinitis with eosinophils | Wood lung or pneumonitis |
| Non-allergic rhinitis with eosinophils | Irritant-induced rhinitis | Silo-filler’s disease |
| Irritant-induced rhinitis | Reactive upper airways dysfunction syndrome | Aspergillus-induced allergic or hypersensitivity conditions |
| Reactive upper airways dysfunction syndrome | Idiopathic rhinitis | Allergic Aspergillus rhinosinusitis |
| Idiopathic rhinitis | Chronic rhinosinusitis | Maltworker lung |
| Chronic rhinosinusitis | Chronic maxillary sinusitis | Allergic bronchopulmonary aspergillosis |
| Chronic maxillary sinusitis | Chronic frontal sinusitis | Allergic or hypersensitivity disorders involving the eye |
| Chronic frontal sinusitis | Chronic ethmoidal sinusitis | Allergic conjunctivitis |
| Chronic ethmoidal sinusitis | Chronic sphenoidal sinusitis | Vernal keratoconjunctivitis |
| Chronic sphenoidal sinusitis | Chronic pansinusitis | Giant papillary conjunctivitis |
| Chronic pansinusitis | Samter’s syndrome | Atopic keratoconjunctivitis |
| Samter’s syndrome | Incl.: Widal Syndrome, Widal Triad | Allergic conjunctivitis due to drugs and medicaments |
| Allergic Aspergillus rhinosinusitis | | Irritant contact blepharoconjunctivitis |

### Asthma

- Allergic asthma
- Non-allergic asthma
- Other and unspecified asthma
- Other specified asthma
- Aspirin-induced asthma
- Exercise-induced bronchospasm
- Cough variant asthma
- Other asthma
- Samter’s syndrome
- Unspecified asthma
- Unspecified asthma with exacerbation
- Unspecified asthma with status asthmaticus
- Unspecified asthma, uncomplicated

### Drug-induced bronchospasm

- Bronchospasm provoked by allergy to food substance

### Hypersensitivity Pneumonitis

- Hypersensitivity pneumonitis due to specific organic dust
- Farmer lung
- Bagassosis

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### Table. Continued

| **Atopic eczema; photoaggravated** | **Chronic urticaria of undetermined aetiology** |
| **Atopic xeroderma** | **Contact urticaria** |
| **Allergy to substances in contact with the skin** | **Allergic contact urticaria** |
| **Allergic contact dermatitis** | **Allergic contact urticaria: localized** |
| Occupational allergic contact dermatitis | **Allergic contact urticaria: disseminated** |
| Allergic contact dermatitis due substantially to occupational exposure to allergen | **Oral urticaria syndrome** |
| Allergic contact dermatitis due in part to occupational exposure to allergen | **Occupational allergic contact urticaria** |
| Allergic contact dermatitis organized by allergen class (covers 17 entities) | **Contact urticaria due to food allergen** |
| Allergic contact dermatitis organized by site (covers 12 entities) | **Non-allergic contact urticaria** |
| **Photo-allergic contact dermatitis** | **Occupational non-allergic contact urticaria** |
| Photo-allergic contact dermatitis organized by photo-allergen class (covers 6 entities) | **Physical urticaria and angioedema** |
| Occupational photo-allergic contact dermatitis | **Dermographism** |
| **Allergic contact urticaria** | **Cold urticaria** |
| Allergic contact urticaria: localized | **Heat contact urticaria** |
| Allergic contact urticaria: disseminated | **Vibratory angioedema** |
| Oral allergy syndrome | **Solar urticaria** |
| Occupational allergic contact urticarial | **Cholinergic urticarial and related conditions** |
| Contact urticarial due to food allergen | **Cholinergic urticaria** |
| **Protein contact dermatitis** | **Cholinergic pruritus** |
| Protein contact dermatitis due to plant protein | **Cholinergic erythema** |
| Protein contact dermatitis due to animal protein | **Exercise-induced anaphylaxis** |
| Occupational protein contact dermatitis | **Food-dependent exercise-induced anaphylaxis** |
| Exacerbation of constitutional dermatitis due to exposure to contact allergens | **Miscellaneous urticarial disorders** |
| Cutaneous reactions to systemic exposure to contact allergens | **Aquagenic urticaria** |
| Systemic contact dermatitis due to ingested allergen | **Angioedema** |
| Symmetrical drug-related intertriginous and flexural erythema | **Urticaria** |
| Systemic contact dermatitis due to implanted allergen | **Urticarial vasculitis** |
| Certain specified allergic reactions to substances in contact with skin and mucous membranes | **Anaphylaxis due to radiocontrast media** |
| **Allergic contact sensitization** | **Syndromes with urticarial reactions or angioedema** |
| **Allergic contact sensitization organized by allergen class (covers 15 entities)** | **Cryopyrin-associated periodic syndromes** |
| **Allergic contact sensitization organized by allergen class (covers 15 entities)** | **Schnitzler syndrome** |
| **Allergic contact sensitization organized by occupational exposure to allergen** | **Episodic angioedema with eosinophilia** |
| **Urticaria, angioedema and other urticarial disorders** | **Tumour necrosis factor receptor 1 associated periodic syndrome** |
| **Spontaneous urticaria** | **Angioedema due to disordered complement** |
| Acute urticaria | **Hereditary angioedema** |
| Acute urticaria due to IgE-mediated allergy | **Hereditary angioedema type I** |
| Acute urticaria due to underlying infection or infestation | **Hereditary angioedema type II** |
| Acute urticaria due to pseudoallergy | **Hereditary angioedema type III** |
| Acute urticaria due to other specified mechanism | **Acquired angioedema** |
| Acute urticaria of undetermined aetiology | **Acquired angioedema type I** |
| **Chronic urticaria** | **Acquired angioedema type II** |
| Chronic autoimmune urticaria | **Drug-induced urticaria** |
| Chronic urticaria due to underlying infection or infestation | **Drug-induced angioedema** |
| Chronic urticaria due to pseudoallergy | **Angioedema due to angiotensin converting enzyme inhibitor** |
| Chronic urticaria due to other specified mechanism | **Drug-induced anaphylaxis** |

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### Anaphylaxis

Anaphylaxis classified by clinical severity (extension codes)
- Anaphylaxis grade 1 [single system]
- Anaphylaxis grade 2 [more than one system; not life-threatening]
- Anaphylaxis grade 3 [more than one system; life-threatening]
- Anaphylaxis grade 4 [life-threatening with cardiac arrest]

Anaphylaxis due to allergic reaction to food
- Food-dependent exercise-induced anaphylaxis
- Drug-induced anaphylaxis
- Anaphylaxis due to insect venom
- Anaphylaxis due to inhaled allergens
- Anaphylaxis due to contact with allergens
- Anaphylaxis provoked by physical factors
  - Cold-induced anaphylaxis
  - Exercise-induced anaphylaxis
  - Food-dependent exercise-induced anaphylaxis
- Anaphylaxis secondary to mast cell disorder

### Complex hypersensitivity/allergic disorders

**Drug Hypersensitivity**

**Drug eruptions**
- Exanathemoid drug eruption
- Drug-induced urticaria
- Drug-induced angioedema
- Fixed drug eruption
  - Limited fixed drug eruption
  - Generalized fixed drug eruption
- Allergic contact dermatitis due to topical medicaments
- Allergic contact dermatitis due to systemic medicaments
- Eczematous drug eruption
- Lichenoid drug eruption
- Stevens-Johnson syndrome and toxic epidermal necrolysis due to drug
  - Drug-induced Stevens-Johnson syndrome
  - Drug-induced Toxic Epidermal Necrolysis
  - Drug-induced Stevens-Johnson syndrome/Toxic Epidermal Necrolysis
- DRESS syndrome
- Acneform and pustular eruptions due to drug
- Drug-associated immune complex vasculitis
- Drug-induced erythroderma
- Drug-induced erythema nodosum
- Miscellaneous specified cutaneous eruptions due to drugs

**Specific organ or system reaction due to drug hypersensitivity**
- Drug-associated immune complex arthritis
- Drug-induced aplastic anemia
- Drug-induced liver hypersensitivity disease
- Drug-induced cytopenia
- Drug-induced bronchospasm

### Drug-induced rhinitis
- Allergic conjunctivitis due to drugs and medicaments
- Drug-induced vasculitis
- Aspirin-induced asthma
- Samter’s syndrome
  - Multiple drug hypersensitivity syndrome

**Food hypersensitivity**
- Food-induced urticarial or angioedema
- Contact urticarial due to food allergen
- Anaphylaxis due to allergic reaction to food
  - Food-dependent exercise-induced anaphylaxis
- Bronchospasm provoked by allergy to food substance
- Oral allergy syndrome
- Allergic contact dermatitis due to food allergen
- Food-induced gastrointestinal hypersensitivity
- Food-induced eosinophilic gastroenteritis
- Food-induced eosinophilic oesophagitis
- Allergic and dietetic colitis
  - Food-induced proctocolitis or colitis of infants
- Allergic and dietetic enteritis of small intestine
  - IgE-mediated allergic enteritis of small intestine
  - Eosinophilic enteritis
  - Food-protein induced enterocolitis syndrome

**Hymenoptera and other insects hypersensitivity or allergy**
- Systemic allergic reaction due to Hymenoptera venom
- Anaphylaxis due to insect venom
- Cutaneous reactions to Hymenoptera venom
- Cutaneous reactions to arthropods
  - Insect bites and stings (covers 9 entities)
  - Arachnid bites and stings (covers 7 entities)
  - Other cutaneous reactions to arthropods

**Allergic or hypersensitivity disorders involving the gastrointestinal tract**

**Allergic gastritis**
- Allergic gastritis due to IgE-mediated hypersensitivity
  - Food-induced IgE-mediated gastrointestinal hypersensitivity
- Allergic gastritis due to non-IgE-mediated hypersensitivity
  - Food-induced non-IgE-mediated gastrointestinal hypersensitivity
- Allergic duodenitis
- Allergic and dietetic colitis
- Food-induced proctitis or colitis of infants
- Food-induced eosinophilic gastroenteritis
- Food-induced eosinophilic oesophagitis
- Allergic and dietetic enteritis of small intestine
  - IgE-mediated allergic enteritis of small intestine
  - Eosinophilic enteritis
  - Food-protein induced enterocolitis syndrome
tuned according to further implementations and adaptations; however, we believe that the ICD revision innovative process, allowing stakeholders to be involved, is critical to increases in the acceptability as well as the accuracy of use of this classification system. This opened and transparent transition allows conditions previously invisible or undernotified to be well classified, which will change dramatically the landscape in which the health system operates.

Currently, we are unable to objectively measure the consequences of these changes in the ICD framework, but we strongly believe that the outcomes of all past and future actions will impact positively as an aggregate data to perform positive quality-improvement in health professional clinical practice as well as can contribute to strengthening the identity of the allergy specialty.

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