



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

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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

INTRODUCTION

Allergy and hypersensitivity, originally perceived as rare and secondary disorders, are one of the fastest growing conditions worldwide becoming a major public health problem, and numerous reports over the last 20 years have been indicating that the world is dealing with an allergy epidemic. They can be expressed in many different organs, with variability of severity degrees, and in any age group, having a significant impact on the quality of life of patients and their families.^{1,2} Every health professional can face them; however, they cannot be adequately tracked by international health classification and coding systems, such as the International Classification of Diseases (ICD). As an example, in 2012, we confirmed the under notification of anaphylaxis deaths due to difficult coding under the ICD-10 using the Brazilian national database,³ which impacts the lack of epidemiological data to support public and private decision-making to offer appropriate treatment, such as auto-injectable adrenaline, still missing in some countries. Apart from mortali-

ty data, morbidity investigations are also likely to be affected by the difficult ICD coding of these conditions. The following publications drew attention to the inadequacy of the ICD-10 (2010 version) and ICD-11 (May 2014 version) frameworks for recording all allergic and hypersensitivity conditions.⁴

The ICD is a key instrument of the World Health Organization (WHO) and a member of the WHO Family of International Classifications (WHO-FIC), which seeks to provide a public global standard to organize and classify information about diseases and related health problems. If the records are unable to

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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.⁵

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-offs⁴ 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.⁶ The high level complex structure underwent a cross-linking terms process to contribute to aligning the allergic and hypersensitivity conditions classification to the ICD-11 beta draft facilitating the classification proposal acceptance.⁷

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 classifica-

tion ICD-11 beta draft facilitating its acceptance.⁶ 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 framework⁸ 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 (IUIS). 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^{6,8}**Allergic or hypersensitivity disorders involving the respiratory tract****Allergic and non-allergic rhinitis**

Allergic rhinitis

Allergic rhinitis due to allergens

Allergic rhinitis due to pollen

Allergic rhinitis due to house dust mite

Other allergic rhinitis due to allergens

Other allergic rhinitis

Non-allergic rhinitis

Gustatory rhinitis

Hormonal-induced rhinitis

Rhinitis related to pregnancy

Rhinitis related to hypothyroidism

Drug-induced rhinitis

Non-allergic rhinitis with eosinophils

Irritant Induced-rhinitis

Reactive upper airways dysfunction syndrome

Idiopathic rhinitis

Chronic rhinosinusitis

Chronic maxillary sinusitis

Chronic frontal sinusitis

Chronic ethmoidal sinusitis

Chronic sphenoidal sinusitis

Chronic pansinusitis

Samter's syndrome

Incl.: Widal Syndrome, Widal Triad

Allergic Aspergillus rhinosinusitis

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

Bird fancier lung

Suberosis

Maltworker lung

Mushroom-worker lung

Maple-bark-stripper lung

Air-conditioner and humidifier lung

Cheese washer's lung

Coffee worker's lung

Fishmeal worker's lung

Grainhandler's disease or lung

Pituitary-snuff-taker's disease

Red-cedar lung or pneumonitis

Wood lung or pneumonitis

Silo-filler's disease

Aspergillus-induced allergic or hypersensitivity conditions

Allergic Aspergillus rhinosinusitis

Maltworker lung

Allergic bronchopulmonary aspergillosis

Allergic or hypersensitivity disorders involving the eye

Allergic conjunctivitis

Vernal keratoconjunctivitis

Giant papillary conjunctivitis

Atopic keratoconjunctivitis

Allergic conjunctivitis due to drugs and medicaments

Irritant contact blepharconjunctivitis

Allergic or hypersensitivity disorders involving skin and mucous membranes**Atopic eczema**

Infantile atopic eczema

Infantile atopic eczema: impetiginization

Childhood atopic eczema

Childhood atopic eczema: flexural

Childhood atopic eczema: nummular pattern

Childhood atopic eczema: chronic lichenified

Childhood atopic eczema: generalized erythematous

Childhood atopic eczema: prurigo pattern

Childhood atopic eczema: impetiginization

Adult atopic eczema

Adult atopic eczema: flexural

Adult atopic eczema: nummular pattern

Adult atopic eczema: chronic lichenified

Adult atopic eczema: generalized erythematous

Adult atopic eczema: prurigo pattern

Adult atopic eczema: impetiginization

Atopic eczema: special forms

Atopic cheilitis

Atopic eczema of eyelids

Atopic eczema of the hands

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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 due to occupational exposure to allergen	Schnitzler syndrome
Urticaria, angioedema and other urticarial disorders	Episodic angioedema with eosinophilia
Spontaneous urticaria	Tumour necrosis factor receptor 1 associated periodic syndrome
Acute urticaria	Angioedema due to disordered complement
Acute urticaria due to IgE-mediated allergy	Hereditary angioedema
Acute urticaria due to underlying infection or infestation	Hereditary angioedema type I
Acute urticaria due to pseudoallergy	Hereditary angioedema type II
Acute urticaria due to other specified mechanism	Hereditary angioedema type III
Acute urticaria of undetermined aetiology	Acquired angioedema
Chronic urticaria	Acquired angioedema type I
Chronic autoimmune urticaria	Acquired angioedema type II
Chronic urticaria due to underlying infection or infestation	Drug-induced urticarial, angioedema and anaphylaxis
Chronic urticaria due to pseudoallergy	Drug-induced urticaria
Chronic urticaria due to other specified mechanism	Drug-induced angioedema
	Angioedema due to angiotensin converting enzyme inhibitor
	Drug-induced anaphylaxis

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Table. Continued

Anaphylaxis	Drug-induced rhinitis
Anaphylaxis classified by clinical severity (extension codes)	Allergic conjunctivitis due to drugs and medicaments
Anaphylaxis grade 1 [single system]	Drug-induced vasculitis
Anaphylaxis grade 2 [more than one system; not life-threatening]	Aspirin-induced asthma
Anaphylaxis grade 3 [more than one system; life-threatening]	Samter's syndrome
Anaphylaxis grade 4 [life-threatening with cardiac arrest]	Multiple drug hypersensitivity syndrome
Anaphylaxis due to allergic reaction to food	Food hypersensitivity
Food-dependent exercise-induced anaphylaxis	Food-induced urticarial or angioedema
Drug-induced anaphylaxis	Contact urticarial due to food allergen
Anaphylaxis due to insect venom	Anaphylaxis due to allergic reaction to food
Anaphylaxis due to inhaled allergens	Food-dependent exercise-induced anaphylaxis
Anaphylaxis due to contact with allergens	Bronchospasm provoked by allergy to food substance
Anaphylaxis provoked by physical factors	Oral allergy syndrome
Cold-induced anaphylaxis	Allergic contact dermatitis due to food allergen
Exercise-induced anaphylaxis	Food-induced gastrointestinal hypersensitivity
Food-dependent exercise-induced anaphylaxis	Food-induced eosinophilic gastroenteritis
Anaphylaxis secondary to mast cell disorder	Food-induced eosinophilic oesophagitis
Complex hypersensitivity/allergic disorders	Allergic and dietetic colitis
Drug Hypersensitivity	Food-induced proctocolitis or colitis of infants
Drug eruptions	Allergic and dietetic enteritis of small intestine
Exanthematic drug eruption	IgE-mediated allergic enteritis of small intestine
Drug-induced urticaria	Eosinophilic enteritis
Drug-induced angioedema	Food-protein induced enterocolitis syndrome
Fixed drug eruption	Hymenoptera and other insects hypersensitivity or allergy
Limited fixed drug eruption	Systemic allergic reaction due to Hymenoptera venom
Generalized fixed drug eruption	Anaphylaxis due to insect venom
Allergic contact dermatitis due to topical medicaments	Cutaneous reactions to Hymenoptera venom
Allergic contact dermatitis due to systemic medicaments	Cutaneous reactions to arthropods
Eczematous drug eruption	Insect bites and stings (covers 9 entities)
Lichenoid drug eruption	Arachnid bites and stings (covers 7 entities)
Stevens-Johnson syndrome and toxic epidermal necrolysis due to drug	Other cutaneous reactions to arthropods
Drug-induced Stevens-Johnson syndrome	Allergic or hypersensitivity disorders involving the gastrointestinal tract
Drug-induced Toxic Epidermal Necrolysis	Allergic gastritis
Drug-induced Stevens-Johnson syndrome/Toxic Epidermal Necrolysis	Allergic gastritis due to IgE-mediated hypersensitivity
DRESS syndrome	Food-induced IgE-mediated gastrointestinal hypersensitivity
Acneform and pustular eruptions due to drug	Allergic gastritis due to non-IgE-mediated hypersensitivity
Drug-associated immune complex vasculitis	Food-induced non-IgE-mediated gastrointestinal hypersensitivity
Drug-induced erythroderma	Allergic duodenitis
Drug-induced erythema nodosum	Allergic and dietetic colitis
Miscellaneous specified cutaneous eruptions due to drugs	Food-induced proctitis or colitis of infants
Specific organ or system reaction due to drug hypersensitivity	Food-induced eosinophilic gastroenteritis
Drug-associated immune-complex arthritis	Food-induced eosinophilic oesophagitis
Drug-induced aplastic anaemia	Allergic and dietetic enteritis of small intestine
Drug-induced liver hypersensitivity disease	IgE-mediated allergic enteritis of small intestine
Drug-induced cytopenia	Eosinophilic enteritis
Drug-induced bronchospasm	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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