

# Coding of allergic diseases in the new version International Classification of Diseases ICD-11n

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## Introduction

The International Statistical Classification of Diseases (ICD) is a classification system created and owned by the World Health Organization (WHO). The history of the ICD goes back over 100 years. The first version of the so-called "International List of Causes of Death" was created in the eighteenth century (1893) and concerned only the causes of death. The subsequent updates were extended to include numerous disease entities and health problems, up to the current tenth version of the ICD-10, approved in May 1990. The ICD has been translated into more than 40 languages and is used in more than 150 countries. The International Classification of Diseases is updated regularly, averaged every 10 years. ICD-10 is widely used for the reimbursement and deployment of funds in the healthcare system.<sup>[1]</sup> ICD-10 has been in force in Poland since 1997. The procedure coding is used to estimate the costs of performed procedures, including surgical interventions.<sup>[2]</sup> The National Health Fund decides to grant reimbursement for a specific treatment, based on ICD codes, which are attached to invoices issued by the treatment centres. Each procedure performed on a patient during hospitalization is coded. If a patient needs more than one procedure, the treatment procedures (e.g. surgical intervention) have more beneficial coding than diagnostic procedures, and thus are better paid by the National Health Fund. The current version of the ICD is not sufficient enough for the precise verification of the procedure payments and the assignment of patients to specific therapeutic groups, therefore it needs to be extended. Based on numerous analyses, it has been shown that the ICD has become an important tool in the field of health, including monitoring disease prevalence, assessing causes of death, external causes of diseases, including drug resistance and monitoring rare diseases.

Some countries, including Australia, Germany, Canada and the USA have expanded the ICD-10 by adapting it to their own needs. With time and the rapid development of individual fields of medicine, the need for detailed recording and documentation of the new clinical conditions has increased, therefore the previous version began to lose its relevance. The work on ICD-11 began in 2011, in June 2018 the World Health Organization (WHO) published on its website a new version of the International Statistical Classification of Diseases and Related Health Problems). In May 2019, the ICD-11 was approved by the 72nd World Health Assembly (WHA72). Currently, the version is being consulted with the WHO member countries and enters into common use on 1 January 2022. The main reason of creation the new ICD-11, was the need to reorganize all diseases basing on the latest medical knowledge. The study involved not only doctors but also public health professionals, statisticians, epidemiologists and representatives of institutions financing public health systems.<sup>[1]</sup>

The ICD-11 classification has been modified and adapted for clinical purposes, for statistics, monitoring morbidity and mortality and will also be a helpful tool for planning expenses in the health care system. It is worth noting, that more than 70% of global health expenditure is currently based, according to the National Centre for Health Statistics, on the ICD classification, which in 2018 accounted for \$3.6 trillion.<sup>[3]</sup> The complete digitalization of the latest version of the ICD will be an event of the 21st century. The fully electronic version will significantly facilitate the work with the IT systems in health care facilities.

## Differences between ICD-10 and ICD-11

In comparison to the previous version of ICD-10, ICD-11 will be more precise and transparent. The classification will still be based on alphanumeric coding, while the number of chapters, individual characters and possible codes have been significantly increased. The latest version of the ICD will contain approximately 55,000 codes, with the possibility of extending the database. In order to facilitate the coding of individual diseases and medical interventions, completely new chapters have been created. ICD-11 consists of 26 main chapters and 2 additional subsections (V and X), which include systems and individual organs, causes of diseases and deaths. There is also a chapter on traditional medicine. The latest version of the ICD has been extended by family medicine issues, antibiotic resistance, a list of drugs, allergens, chemicals, as well as results of histopathological examination.

The tenth revision uses alphanumeric coding with a letter in the first place, numbers in the next three positions and the fourth character after the period. The codes start from A00.0 till Z99.9. Each nosological unit is marked alphanumerically: A12.X, where A - is a letter, 12 is a number, X - a number that identifies the subtype. The subtype 8 indicates "other" and the subtype 9 indicates "undefined". The "U" codes are intended for the new diseases of uncertain aetiology. As for the 11 version of the ICD, the codes range from 1A00.00 to ZZ9Z.ZZ, while the codes in group "X" are extension codes, that allow a more detailed description of the course of the disease. For example, allergic asthma which in ICD-10 is under the code of J45.0, in ICD-11 is under the code of CA23.0. Extended ICD-11 database, enables us to code the allergic, severe asthma with acute episodes: CA23.00 & XS25. The basic differences between ICD-10 and ICD-11 are presented in Table 1.

ICD version	ICD-10	ICD-11
Number of codes	14.4 thousand	55 thousand
Chapters	21 chapters	26 chapters, 2 subsections
Alphanumeric coding	A00.0-Z99.9	1A00.00-ZZ9Z.ZZ
Possibility to extend the code	No	Yes, codes from the "X" group

## Allergic diseases in ICD-11

Allergic diseases are considered to be the epidemic of the 21st century. It is estimated that almost one third of the world's population (25-30%) suffers from allergic diseases, including over 150 million Europeans. and the figures are rising drastically every year.<sup>[4]</sup> Allergy and hypersensitivity are the fourth most common group of chronic diseases and one of the main problems in public health. The WHO predicts that in 2050 every second person in the world will suffer from allergy, especially in industrialized countries. Due to the multi-organ clinical manifestation of allergic diseases, specialists in many fields of medicine have contact with allergic patients. Most cases of allergies or hypersensitivity are treated by general practitioners, paediatricians, ERT (Emergency Rescue Team) and UCU (Urgent Care Unit) physicians.

**In the current ICD-10, allergic conditions were characterized too generally.** The allergic diseases include asthma, anaphylaxis, allergic rhinitis, food and drug allergy, urticaria, angioedema, atopic dermatitis and many others. Apart from their wide clinical manifestation, the allergic diseases are characterized by various pathomech-

anisms, triggers and symptom severity. This affects not only the patient's quality life, but also the costs of treatment for patient and the entire healthcare system. The diagnosis of an allergic disease is not always easy and is based mainly on a carefully collected medical history, physical examination, and in vivo and in vitro diagnostic tests. The basic in vivo tests include skin prick tests, intradermal tests - used in the diagnosis of IgE-dependent allergy, patch tests - used in the diagnosis of contact allergy, and provocation tests - the "gold standard" in the diagnosis of food, drug or inhalation allergy. Provocation tests allow to confirm the diagnosis, indicate factors causing an allergic reaction, choose a safe drug, test the new therapeutic options, and monitor the course of the disease.<sup>[5]</sup> It is worth mentioning, that in vivo tests, such as intradermal and patch tests are used in allergology for over 100 years, due to their very high usefulness, which is emphasized by the current guidelines and recommendations of recognized Allergy Scientific Societies<sup>[5,6]</sup> On the other hand, in vitro studies such as determination of specific IgE concentration, tryptase level, basophil activation tests (BAT), lymphocyte transformation tests (LTT) are helpful in confirming the diagnosis and are a safe alternative to in vivo tests, especially among patients with severe allergic diseases.<sup>[7]</sup> These tests cannot be used as screening tests and their results should always be interpreted with caution. Most of these methods still require validation to ensure appropriate sensitivity and specificity. Proper diagnostics, both in vivo and in vitro, is helpful in optimizing the treatment expenses in both health care systems (public and private). In the current model of the health care system, the costs of diagnosis and treatment of a patient are based on the coding of specific diseases. In this regard, the ICD is the most common system that tracks the incidence of diseases, the costs of their diagnosis and treatment.

So far, the allergic diseases have not been accurately represented and properly coded in the ICD. This resulted in underestimation of their occurrence and errors in statistical studies on incidence and mortality as well as had a negative impact on the diagnosis and treatment of patients. In 2012, Tanno et al. presented data according to which the ICD-10 does not reliably provide the number of deaths caused by anaphylaxis, significantly underestimating this value.<sup>[8]</sup> It soon turned out that the problem also refers to other allergic diseases and hypersensitivity reactions. In the international survey on classification and coding of allergic diseases, which was attended by 612 EAACI and/or WAO members from 144 countries, the majority of respondents considered that the ICD, despite being the most common disease classification system in the world, is not suitable for use in clinical practice. According to most of the respondents, the classifying of hypersensitivity/allergic diseases in the ICD-10 is neither easy nor accurate.<sup>[9]</sup> The work on the new ICD

qualification has become a unique opportunity to improve the classification of allergic disease coding. It was necessary to prove the need to make changes in the ICD, with respect to the allergic diseases and prepare an appropriate high-quality document for WHO ready for implementation in the new ICD-11 edition.

The hardest thing in formation of the new ICD-11 classification was to create appropriate codes for diseases in connection with the diagnostic procedures necessary for the correct diagnosis.

Under the leadership of Luciana Tanno and Pascal Demoly, a list of diagnostic procedures (keywords) relevant for allergic/hypersensitivity diseases and used by specialists in their daily practice was prepared. (Table 2).

**Table 2** Diagnostic procedures used in allergology.<sup>[10]</sup>

The authors of the study were aware of the fact, that some in vitro procedures have been used so far only for the purposes of clinical trials, in selected research centres in few countries, but the increasing number of publications on their usefulness suggested that they would be most likely used in everyday practice in the future. Selected procedures in the ICD-10 (2015 version) were compared with the ICD-11 beta version (2015). The analysis of currently applied ICD-10 and the draft ICD-11 version has drawn a picture of many shortcomings, imprecise terminology and the dispersion of allergology issues across the various chapters of the ICD.

Diagnostic procedures for IgE-mediated hypersensitivity	Diagnostic procedures for T cell mediated hypersensitivity
<b>In vitro</b>	<b>In vitro</b>
<ul style="list-style-type: none"> <li>Serum specific IgE</li> <li>tryptase concentration in blood serum (plasma)</li> <li>basophil activation test (BAT)</li> <li>cellular antigen stimulation test (CAST) - enzyme immunoassay (ELISA)</li> </ul>	<ul style="list-style-type: none"> <li>lymphocyte transformation test (LTT)</li> <li>ELISPOT test</li> <li>CD69 expression</li> </ul>
<b>In vivo</b>	<b>In vivo</b>
<ul style="list-style-type: none"> <li>skin tests</li> <li>skin prick tests</li> <li>intradermal tests</li> <li>provocation tests</li> </ul>	<ul style="list-style-type: none"> <li>skin tests</li> <li>skin prick tests and/or photopatch tests</li> <li>intradermal tests</li> <li>provocation tests</li> </ul>

The authors proposed a clear presentation of the procedures used in the diagnosis of allergic diseases/hypersensitivity in a separate group, and at the same time their

update it in the International Classification of Health Interventions (ICHI).<sup>[10]</sup>

Joint forces of the European Academy of Allergy and Clinical Immunology (EAACI), World Allergy Organization (WAO), American Academy of Allergy Asthma and Immunology (AAAAI), Latin American Allergy, Asthma and Immunology Association (SLAAI), Asia Pacific Association of Allergy, Asthma and Clinical Immunology (APAAACI) and the American College of Allergy, Asthma and Immunology (ACAAI) created the "Joint Allergy Academies". Thanks to international cooperation of allergy societies, an extensive two-page list of allergic diseases/hypersensitivity was presented, which, after consultation with the WHO, was included in the draft ICD- 11.<sup>[11]</sup> Over the past seven years, a series of publications have been released, so that for the first time in the history of the ICD, allergic/hypersensitivity diseases had a completely separate chapter.<sup>[12]</sup> The enormous amount of work done by Luciane Kase Tanno and Pascal Demoly resulted in the creation of the WHO Collaborating Centre WHO CC (2018) for the Scientific Classification of Allergic Diseases and Hypersensitivity at the Department of Pulmonology and Allergology of the University Hospital of Montpellier.<sup>[13]</sup> WHO CC Montpellier is one of 25 WHO centres which collaborates in the international classifications and the only centre responsible for the allergic diseases. The WHO Montpellier Centre will be involved in promoting the implementation, workers training, application and updating of ICD classification to improve the quality of allergic diseases coding. The latest version of ICD-11 is available in English on the WHO official website.<sup>[14]</sup> Intense work of the WHO CC resulted in the creation of a separate section titled: Allergic and hypersensitivity conditions in the newly created chapter *Disorders of the Immune System* in ICD-11.<sup>[15]</sup>

The new rules for classifying and characterizing allergic diseases in the ICD-11 have undergone extensive functional and quality tests, which have confirmed the increased accuracy of allergy and hypersensitivity coding compared to the ICD-10.<sup>[16]</sup> ICD-11 allows the diagnoses to be linked to a number of additional parameters by adding one or more extensions to the main code. The extensions are listed in Chapter X under the name of "Extension Codes". The WHO supported such a procedure, that will enable the appropriate linking of apparently different but important values for the course of allergic diseases, such as clinical symptoms severity, location or the cause of the disease. The WHO classification of death cause codes has also been revised by including anaphylaxis in the list of official causes of death, which will allow in future to include anaphylaxis in the official death records. This will improve the precision of mortality statistics and the analysis of related factors, e.g. aetiology.<sup>[16]</sup>

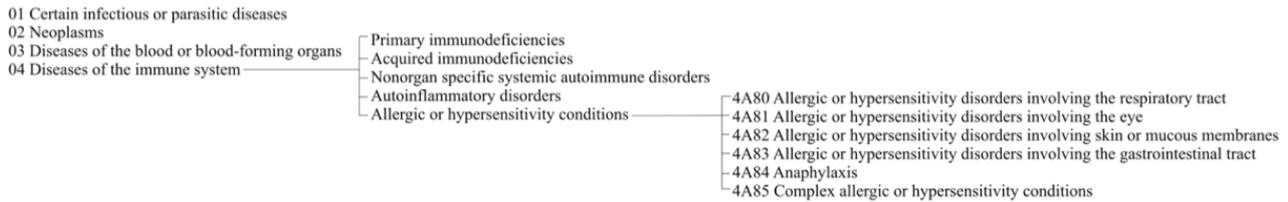


Figure 1. ICD-11, chapter 4 - classification concerning allergic diseases.<sup>[14]</sup>

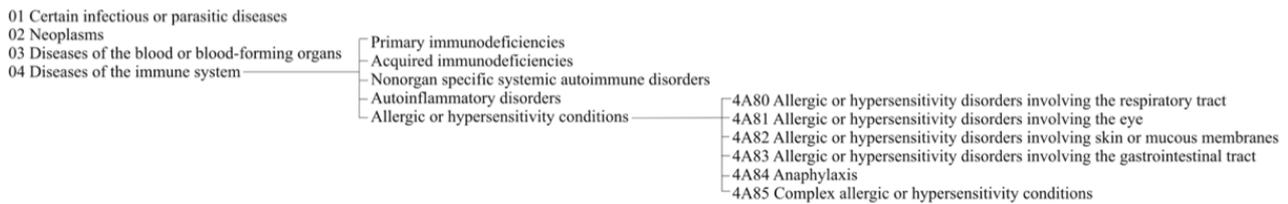


Figure 1. Allergens listed in ICD-11 chapter X.<sup>[14]</sup>

### ICD-11 chapter 4 "Allergic diseases and hypersensitivity"

Due to the various clinical manifestation of allergic diseases, a completely new subchapter entitled "Allergic or hypersensitivity conditions" was created in the fourth chapter entitled "Diseases of immune system". In this chapter issues related to allergy/hypersensitivity (A/H) were divided into individual groups, and these, in turn, into subgroups that define specific diseases (Figure 1). Each of the subgroups has been significantly expanded, which allowed to assign the most common allergic diseases to a specific code, including rare diseases such as hereditary angioedema -HAE (4A00.1).

Allergic diseases have the codes of chapter four, but in this chapter we can also find diseases encoded in other chapters, like spontaneous urticaria, which can be found in subsection 4A82, but has the code EB00.

The new version of ICD also includes codes for „skin or other sensitization tests" QA00.A and for allergen immunotherapy QA00. Thanks to the chapter "X", it is possible to extend the code with the severity of symptoms, the location of changes, as well as a specific allergen (Figure 2.).

A particular attention was paid to the issue of anaphylax-

is. In ICD-10, the anaphylaxis is included in Section T78, " Adverse effects, not elsewhere classified ", of the Chapter XIX, " Injury, poisoning and certain other consequences of external causes." In the updated version, the section on anaphylaxis (4A84) is significantly expanded , and include the types of anaphylaxis, the severity of the course and along with the indication of the causative agent..

Despite the significantly extensive database, the ICD-11 encoding seems to be more intuitive in comparison to the ICD-10. Using the new qualification will not require memorizing individual codes of diseases. For example, by entering the search terms "allergy" and "venom" , the full name of the disease with the suitable code will occur - "Allergic reaction to insect venom" - 4A85.3. There is also a possibility to complete the code by the course of the reaction to : systemic- 4A85.30 or cutaneous- 4A85.31. In addition, the expansion of the codes will reduce the current need to use the descriptions of individual disease in medical documentation.

The ICD let us to follow the occurrence of the diseases, diagnose them, and also support the decision-making of the organizational health care systems in many countries, therefore any changes within the ICD are of great economic importance. Appropriate usage of codes for record-

ing disorders is a key issue to prevent misclassification. The current ICD-10 classification has many limitations. The new, extended version ICD-11 will have a number of positive effects. More reliable classification will improve the quality of statistical and epidemiological analysis of allergic and hypersensitivity diseases. Increase awareness of allergy and improve clinical management of allergic patients. It also significantly improve the work of medical specialists and enable better financing of health services.

**Conflict of interest: none declared.**

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