

# New Histopathological Diagnostic Terms and their Codes of the Central Nervous System (CNS) in the International Classification of Diseases for Oncology (ICD-O 3.1) and about the Turkish Version of ICD-O

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## Abstract

International Classification of Diseases for Oncology (ICD-O), which has been used in the world since 1976 for 40 years and has been translated into many languages, emphasizing the importance of health and cancer is presented. In addition, the histopathological classification working group of central nervous system (CNS) tumors has proposed new terms and their new ICD-O codes, taking into account their genetic characteristics. Our goal is to emphasize the current importance of ICD-O. Since 2015, ICD-O has been put into practice by the Ministry of Health in Turkey. Writing of ICD-O codes is required for pathology reports. In the last 25 years, I have translated the ICD-O three times into Turkish, 1992, 2002 and 2015, serving as pathology, oncology, cancer registrars and the Ministry of Health.

**Keywords:** CNS; New histopathological terms; Neuropathology; ICD-O; ICD-O 3.1; Codes; Turkish version of ICD-O

## Introduction

The International Classification of Diseases for Oncology (ICD-O) has been used for nearly 40 years as a standard tool for coding the site and the histology of the diagnostic terms of the neoplasms in tumor and cancer registrars and in pathology departments and pathology laboratories, usually obtained from a pathology report [1-4].

The ICD-O is the gateway to the world that the latest information has been freely available to entire world's people so that World Health Organization's (WHO) diagnosis and treatment of health and cancer in the world can be made at the highest level of knowledge and scientific benefit.

ICD-O is a dual classification with coding systems for both topography and morphology.

The topography code describes the localization of the tumor and uses the same 3-character and 4-character categories as in the neoplasm section of Chapter II, ICD-10 [5].

The morphology or histopathology code describes the features of the tumor itself, including its cell type and biologic activity and genetic properties.

In preparing the revised edition of the ICD-O 3.1, the editors have made a special effort to change as few terms as possible, to add new terms at empty spaces, and to avoid reuse of previously assigned codes [4]. While all topography or localization codes remain the same as in the previous edition, morphology or histopathology codes have been thoroughly reviewed and, where necessary, revised to increase their diagnostic precision and prognostic value [4].

Currently, in the light of new scientific and technological developments, especially genetic features and characteristics have become very important element in tumor diagnosis.

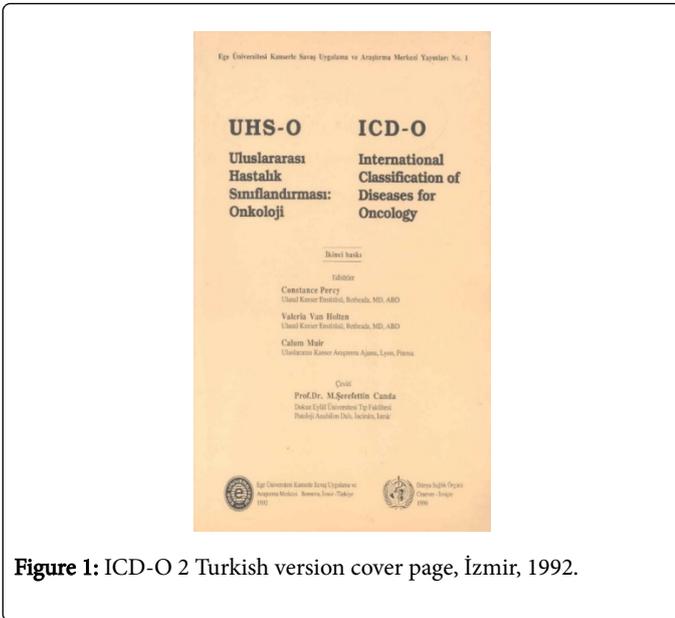
On the other hand, if we look at new information, new technological improvement and new perspectives, the use of the international histopathological classification of tumors and neoplastic diseases has become a compulsory rule to use in pathology in the all over the world. So, this approach is to promote the adoption of a uniform terminology that will facilitate communication and dialog among cancer workers.

As a matter of fact, in the light of new information and scientific developments, the importance of ICD-O 3.1 for this reason it has increased more. Therefore ICD-O books translated many languages around the World including Turkish. I have been working on the Turkish version of ICD-O for the past 25 years since 1991 as reported by Fidaner C et al. [6] and as reported by Eser S et al. [7] and I am very happy to prepare the Turkish version of ICD-O 2, ICD-O 3 and ICD-O 3.1 during this time [8-10] (Figures 1-3).

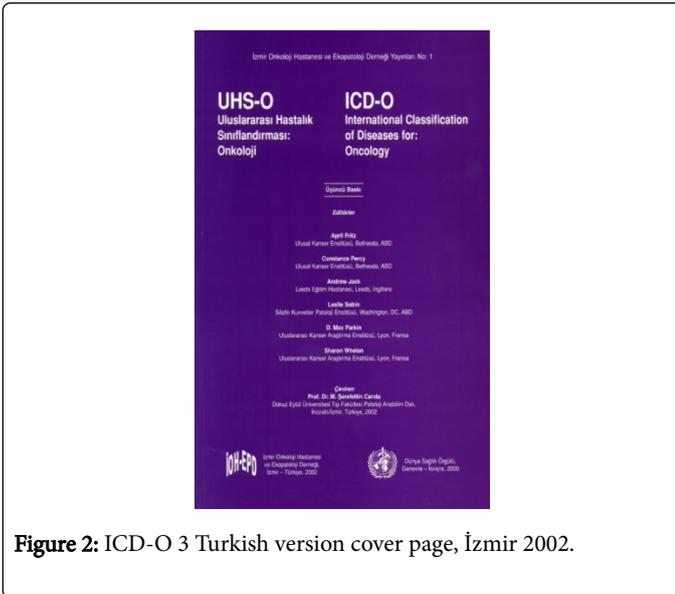
These studies have been made to provide scientific and social benefits as a volunteer and with the sensitivity of a non-profit scientist. Our aim is to get scientific and social benefit in the field of pathology, oncology and cancer registry, besides community health, especially in the fight against cancer in Turkey.

Because ICD-O principles are of contemporary scientific and universal value, on this occasion I must say, it is very important that scientists and healthcare providers apply these principles.

Since the beginning, the Ministry of Health has shown interest in the statistics of cancer patients in Turkey. Ultimately, the use of ICD-O was made mandatory by the Ministry of Health in 2015, in all hospitals, healthcare facilities and university hospitals in Turkey.



**Figure 1:** ICD-O 2 Turkish version cover page, İzmir, 1992.



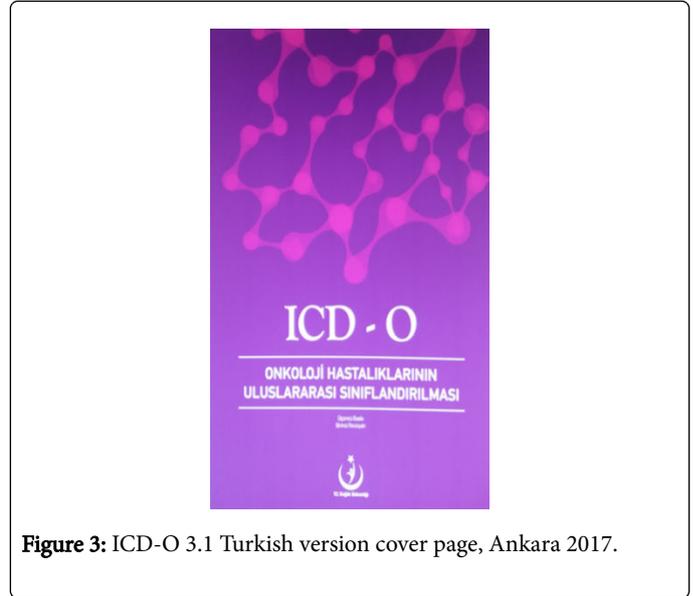
**Figure 2:** ICD-O 3 Turkish version cover page, İzmir 2002.

The decision of the Ministry of Health to implement ICD-O in all health institutions and pathology reports is a valuable and up-to-date scientific development that at the same time opens the door to new scientific and social studies as much as possible to make respectable and scientific cancer statistics in Turkey.

Also, the result of new scientific and technological developments, especially in the following areas, new terms have emerged; Central nervous system (CNS) [11], hematopoietic and lymphoid tissues [12], digestive system [13], breast [14] and urinary system [15] tumors and

so on. In this review new terms and ICD-O codes related to CNS tumors will be presented.

Our aim is to emphasize newly identified tumors, terms and codes in CNS tumors which is seen in the ICD-O 3.1 and to explain as a briefly the progress made about in the use of ICD-O and the Turkish version of ICD-O in Turkey.



**Figure 3:** ICD-O 3.1 Turkish version cover page, Ankara 2017.

### New and Changing Terms of the CNS, and their Codes in ICD-O 3.1

ICD-O 3.1 includes new terms, changing terms, and new codes related to CNS tumors, as in other areas. In particular, the increase in the demand for genetic tests related to histopathology diagnoses and the presence of new technologies make it mandatory [4].

In addition, these new developments and new histopathologic diagnoses make better results on the treatment and prognosis of the disease.

Significant changes have been made in the codes and terms of CNS tumors under the influence of new techniques in use in current histopathology.

Accordingly, ICD-O 3.1 includes new diagnostic and terminology changes as well as concerns about biological behavior changes. These changes have affected the studies related to neuropathology, neurosurgery, neuro-oncology, neuroradiology and neuro-nuclear medicine and have caused many changes in terms of patients in practice.

Therefore, all these new and very important changes specific to CNS tumors, taking advantage of the tables of tumors of all systems in the ICD-O 3.1 original book, as shown below, I also tried to summarize them in tables form [3,4,9,10] (Tables 1-7).

New Codes	Terms
8272/0	Pituitary adenoma, NOS (C75.1)

8272/3	Pituitary carcinoma, NOS (C75.1)
8728/0	Diffuse melanocytosis (C70.9)
8728/1	Meningeal melanocytoma (C70.9)
8728/3	Meningeal melanomatosis (C70.9)
9351/1	Craniopharyngioma, adamantinomatous (C75.2)
9352/1	Craniopharyngioma, papillary (C75.2)
9365/3	Askin tumor
9371/3	Chondroid chordoma
9372/3	Dedifferentiated chordoma
9373/0	Parachordoma
9412/1	Desmoplastic infantile astrocytoma Desmoplastic infantile ganglioglioma
9413/0	Dysembryoplastic neuroepithelial tumor
9444/1	Chordoid glioma (C71._) Chordoid glioma of third ventricle (C71.5)
9474/3	Large cell medulloblastoma (C71.6)
9493/0	Dysplastic gangliocytoma of cerebellum (Lhermitte-Duclos) (C71.6)
9508/3	Atypical teratoid/rhabdoid tumor (C71._)
9571/0	Perineurioma Intraneural perineurioma Soft tissue perineurioma
9571/3	Perineurioma, malignant Perineural MPNST
9582/0	Granular cell tumor of the sellar region (C75.1)

**Table 1:** New codes in ICD-O, third edition (10). (The following 4-digit morphology codes did not exist in ICD-O, second edition.). A term without a number is a synonym for the preceding code.

New Codes	Terms
9362/3	Mixed pineal tumor (C75.3) Mixed pineocytoma-pineoblastoma (C75.3) Pineal parenchymal tumor of intermediate differentiation (C75.3) Transitional pineal tumor (C75.3)
9364/3	Peripheral primitive neuroectodermal tumor, NOS, PPNET
9382/3	Anaplastic oligoastrocytoma (C71._)
9383/1	Mixed subependymoma-ependymoma (C71._)
9390/1	Atypical choroid plexus papilloma (C71.5)
9390/3	Choroid plexus carcinoma (C71.5)
9391/3	Cellular ependymoma (C71._)

	Clear cell ependymoma (C71._) Tancytic ependymoma (C71._)
9400/3	Diffuse astrocytoma (C71._) Astrocytoma, low grade (C71._) Diffuse astrocytoma, low grade (C71._)
9423/3	Polar spongioblastoma (C71._)
9442/1	Gliofibroma (C71._)
9470/3	Melanotic medulloblastoma (C71.6)
9471/3	Desmoplastic nodular medulloblastoma (C71.6)
9473/3	PNET, NOS Central primitive neuroectodermal tumor, NOS (C71._), CPNET (C71._) Supratentorial PNET (C71._)
9500/3	Central neuroblastoma (C71._)
9501/0	Diktyoma, benign (C69._)
9501/3	Diktyoma, malignant (C69._)
9502/0	Teratoid medulloepithelioma, benign (C69.4)
9505/3	Ganglioglioma, anaplastic
9506/1	Central neurocytoma Cerebellar liponeurocytoma Lipomatous medulloblastoma (C71.6) Neurolipocytoma (C71.6) Medulloctoma (C71.6)
9530/3	Meningioma, anaplastic
9538/1	Clear cell meningioma Chordoid meningioma
9538/3	Rhabdoid meningioma
9539/1	Atypical meningioma
9540/3	Malignant peripheral nerve sheath tumor MPNST, NOS MPNST with glandular differentiation Epithelioid MPNST MPNST with mesenchymal differentiation Melanotic MPNST Melanotic psammomatous MPNST
9560/0	Melanotic schwannoma Plexiform schwannoma Cellular schwannoma Degenerated schwannoma Ancient schwannoma Psammomatous schwannoma
9561/3	Malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation MPNST with rhabdomyoblastic differentiation
9560/0	Melanotic schwannoma

	Plexiform schwannoma Cellular schwannoma Degenerated schwannoma Ancient schwannoma Psammomatous schwannoma
9561/3	Malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation MPNST with rhabdomyoblastic differentiation

**Table 2:** New morphology terms and synonyms in ICD-O, third edition (10). (The following 4-digit morphology codes existed in ICD-O, second edition.)

ICD-O, second edition	Term as it appears in ICD-O, third edition	ICD-O, third edition
9422/3	Spongioblastoma, NOS (C71._) [obs]	9421/1
9443/3	Primitive polar spongioblastoma (C71._) [obs]	9423/3
9481/3	Monstrocellular sarcoma (C71._) [obs]	9441/3
9490/0	Gangliocytoma	9492/0
9536/0	Hemangiopericytic meningioma (C70._) [obs]	9150/1
9594/3	Microglioma (C71._) [obs]	9590/3

**Table 3:** Terms that changed morphology code in ICD-O, third edition (10).

	Terms as it appears in ICD-O, third edition	ICD-O, third edition
M-----	Histiocytosis X, NOS	9751/1
M-----	Eosinophilic granuloma	9752/1
M-----	Hand-Schuller-Christian disease	9753/1

**Table 4:** Terms that changed from tumor-like lesions to neoplasms in ICD-O, third edition (10).

ICD-O, second edition	The terms which were deleted for ICD-O, third edition
9382/3	Mixed oligoastrocytoma (replaced with Oligoastrocytoma)
9531/0	Meningothelomatous meningioma (replaced with Meningothelial meningioma)
9560/0	Melanocytic schwannoma (replaced with Melanotic schwannoma)

**Table 5:** Terms in ICD-O, second edition, which were deleted for ICD-O, third edition (10).

ICD-O, second edition	Terms as it appears in ICD-O, third edition	ICD-O, third edition
	Terms Changing from Borderline to Malignant	
9393/1	Papillary ependymoma (C71.)	9393/3
9538/1	Papillary meningioma	9538/3
	Terms Changing from Malignant to Borderline	
9421/3	Pilocytic astrocytoma (C71.)	9421/1
9421/3	Piloid astrocytoma (C71.)	9421/1
9421/3	Juvenile astrocytoma (C71.)	9421/1

9422/3	Spongioblastoma, NOS (C71.) [obs] Terms Changing from Benign to Borderline	9421/1
9506/0	Neurocytoma	9506/1

**Table 6:** New codes, preferred terms, related terms, and synonyms in ICD-O, third edition, first revision (ICD-O 3.1) (10).

Status	ICD-O, 3 Morphology Code	Terms	Action
New term and code	9395/3	Papillary tumor of the pineal region	
New term and code	9425/3	Pilomyxoid astrocytoma	
New term and code	9431/1	Angiocentric glioma	
New term and code	9432/1	Pituicytoma	
New related term	9471/3	Medulloblastoma with extensive nodularity	
New related term	9474/3	Anaplastic medulloblastoma	
New related term	9506/1	Extraventricular neurocytoma	
New term and code	9509/1	Papillary glioneuronal tumor	
New related term	9509/1	Rosette-forming glioneuronal tumor	

**Table 7:** New codes, preferred terms, related terms, and synonyms in ICD-O, third edition, first revision (ICD-O 3.1) (10).

## Conclusion

In the ICD-O 3.1 edition, new histopathologic markers and genetic information were taken into consideration in the new classification of CNS tumors in the field of neuropathology, resulting in the following new developments [4,9,10]:

- New codes,
- New morphology terms and synonyms,
- Terms that changed morphology code,
- Terms that changed from tumor-like lesions to neoplasms,
- Terms which were deleted,
- Terms that changed behavior code,
- New codes, preferred terms, related terms, and synonyms.

Under the influence of the contemporary scientific and technological improvement, the histopathological diagnosis of the patients becomes more realistic, also directs the treatment and have an effect on the prognosis.

As a natural consequence of this improvement, there are new codes and changes in some codes of ICD-O. By the way ICD-O 3.1 was last published by WHO in 2013. In the meantime, in my opinion, the publication of a new edition of the ICD-O has become a necessity. Because under these new circumstances, for example, the result of extraordinary innovations on genetics [16-18] and PET (positron emission tomography) pharmaceuticals [19-21], on the near future, new kind of tumor histopathologic diagnoses can be predicted, so new approaches of pathologists and oncologists will be seen.

These new diagnostic terms and codes mentioned are also important in influencing the patient's prognosis and treatment strategy. For this reason, it should be taken carefully into consideration.

In this study, attention was drawn to new terms, codes and improvement related to CNS tumors, which constitute a special group of human tumors, emphasizing and presenting the importance

At the same time, WHO's ICD-O and Tumor Classification Books and pathological, oncological and coding studies in parallel with these principles is to present comparable data on cancer incidence for all countries around the world for which high-quality data have been made available by population-based cancer registries [22].

As a result, in line with WHO's work, it is a good step for the Ministry of Health to put the ICD-O codes into practice in all hospitals and healthcare facilities, starting in 2015 (2014) in Turkey [23,24].

Similarly, the definition of Pathology reports according to current WHO Tumor Classifications and coding with ICD-O codes is very important and valuable for Turkey in terms of scientific prestige as well as the health and prognosis of patients.

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