



Evropská unie
Evropský sociální fond
Operační program Zaměstnanost



NÁRODNÍ CENTRUM
PRO MEDICÍNSKÉ NOMENKLATURY
A KLASIFIKACE



UNIVERZITA
KARLOVA

Vzácná onkologická onemocnění v ČR z dat Národního onkologického registru

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klasifikace

ÚZIS ČR

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Akce je pořádána v rámci projektu NCMNK - Národní centrum pro medicínské
nomenklatury a klasifikace, registrační číslo
CZ.03.4.74/0.0/0.0/15_025/0016089.

Kla si fi Kon

Konference
o klinických
klasifikačních
a terminologických
systémech
a jejich
použití
v českém
zdravotnictví

Co jsou to vzácné nádory?

Doporučení Rady EU ze dne 8. června 2009 o akci v oblasti vzácných onemocnění:

Členské státy mají používat pro účely politiky jednotnou definici vzácného onemocnění jako **onemocnění, které nepostihuje více než 5 z 10 000 osob (prevalence onemocnění)**.

European Commission. *Rare Diseases*. Available from: https://ec.europa.eu/health/rare_diseases/policy_en.

Definice podle evropské informační sítě RARECARENet:
incidence je < 6 onemocnění na 100 000 osob za rok

- Projekt **Surveillance of Rare Cancers in Europe (RARECARE)** -> Gatta, G., et al., Rare cancers are not so rare: the rare cancer burden in Europe. *Eur J Cancer*, 2011. 47(17): p. 2493-511.
- **RARECARENet**. Cancer List: <http://www.rarecarenet.eu/rarecarenet/index.php/cancerlist>.
- Gatta, G., et al., Burden and centralised treatment in Europe of rare tumours: results of RARECAREnet-a population-based study. *Lancet Oncol*, 2017. 18(8): p. 1022-1039.

Vzácné nádory podle RARECARENet

incidence je < 6 onemocnění na 100 000 osob za rok

Projekt Surveillance of Rare Cancers in Europe (RARECARE)

Gatta, G., et al., Rare cancers are not so rare: the rare cancer burden in Europe.

Eur J Cancer, 2011. 47(17): p. 2493-511.

- European (EU27) patients diagnosed from 1988 to 2002, covered population about 162,000,000
- 108 per 100,000 ~ 541,000 new diagnoses annually ~ **22% of all cancer diagnoses**
- 5-year relative survival worse for rare cancers (47%) than common cancers (65%)
- prevalence about 4,300,000 patients in EU with rare cancer ~ **24% of the total cancer prevalence**

Rare Cancers Europe (RCE, <http://www.rarecancerseurope.org/>) - multi-stakeholder initiative dedicated to putting rare cancers firmly on the European policy agenda and to implementing 39 political and stakeholder recommendations ->

RARECARENet (<http://www.rarecarenet.eu/rarecarenet/>)

Cancer List (Updated October 2015) <http://www.rarecarenet.eu/rarecarenet/index.php/cancerlist>.

Vzácné nádory podle RARECARENet

Cancer List (Updated October 2015) <http://www.rarecarenet.eu/rarecarenet/index.php/cancerlist>.

Rationale

The attached document provides a definition of rare cancers and explains the criteria used for defining the cancer entities shown in the list.

Rationale and Questions for Consensus - [download PDF](#)

List of Rare Cancers

This is the list of tumour entities from which rare tumours are identified as those with incidence less than 6 per 100,000 persons / year. The list presents the number of cases reported by European cancer registries during the period 2000-2007 and the corresponding incidence rates. The list is derived from the data of 94 population-based cancer registries from 24 European countries adhering to the RARECAREnet project.

This list includes information on all cancers defined by the RARECARE and RARECAREnet project. Please note that rare cancers are only those with an incidence of <6/100.000

[Download RARECAREnet List Excel spreadsheet](#) Updated October 2015

Experts for the Definition of Rare Cancers / Rare Cancers List - [download PDF](#)

Rare Cancers List Endorsers

- Childhood Cancer Research Group, <http://www.ccrq.ox.ac.uk>
- CML Advocates Network, <http://www.cmladvocates.net>
- EUROPA DONNA - The European Breast Cancer Coalition: www.europadonna.org
- European Association for Cancer Research (EACR), <http://www.eacr.org>
- European Association of Neuro-Oncology (EANO), <http://www.eano.eu>
- European Association of Nuclear Medicine (EANM), <http://www.eanm.org>
- European Cancer Patient Coalition (ECPC), <http://www.ecpc-online.org>
- European Group for Blood and Marrow Transplantation: www.ebmt.org
- European Oncology Nursing Society, <http://www.cancernurse.eu>
- European Prostate Cancer Coalition, Europa Uomo, <http://www.europa-uomo.org>
- European Society of Gynaecological Oncology (ESGO), <http://www.esgo.org>
- European Society for Medical Oncology (ESMO), <http://www.esmo.org>
- European Society for Paediatric Oncology (SIOP), <http://www.siop.org>
- European Society of Surgical Oncology (ESSO), <http://www.essoweb.org>
- European Society of Therapeutic Radiology and Oncology (ESTRO), <http://www.estro.org>
- International Brain Tumour Alliance (IBTA), <http://www.theibta.org>


Experts for the definition of rare cancers /rare cancers list			
NAME	SURNAME	ORGANISATION	COUNTRY
Eric	Schryvers	Zweverhaasnetwerk Antwerpen ZNA - hospital network	Belgium
Hildrun	Sundseth	European Cancer Patients Coalition (ECPC)	Belgium
Jon	Geissler	European Cancer Patients Coalition (ECPC)	Germany
Jan-Willem	Van De Loo	European Commission	Belgium
Pascalie	Blaes	European Society for Medical Oncology (ESMO)	Belgium
Sandrine	Marraud	European Organisation for Research and Treatment of Cancer (EORTC)	Belgium
Ricardo	Audino	European Society of Surgical Oncology (ESSO)	France
Elien	Benhamou	Institut Gustave Roussy	France
Marine	Grossgoupi	Institut Gustave Roussy	France
Isabelle	Ray-Cogaard	Centre Léon Bérard	France
Jean-Pierre	Droz	Centre Léon Bérard	France
Marc	Maynadie	Cote d'or Haematological Malignancies Registry	France
Séghène	Aymé	The portal for rare diseases and orphan drugs (ORPHANET)	France
Simon	Baconner	Connective tissue cancer network - CORTICANET	France
Markus	Wartenberg	Das Lebenshaus, E.V.-Global GIST Network	Germany
Rüdiger	Hellmann	Leukemia Network	Germany
Sandra	Usody	National Cancer Registry of Ireland	Ireland
Alba	Brandes	Medical Oncology Department, AUSL Bologna	Italy
Alessandro	Grnchi	Fondazione IRCCS, Istituto Nazionale dei Tumori, Milan	Italy
Angelo Paolo	Di Lorenzo	Asenda USL N.3 Regione Veneto-health care unit	Italy
Franco	Bertino	Fondazione IRCCS, Istituto Nazionale dei Tumori, Milan	Italy
Lisa	Licitra	Fondazione IRCCS, Istituto Nazionale dei Tumori, Milan	Italy
Paolo	Casali	Fondazione IRCCS, Istituto Nazionale dei Tumori, Milan	Italy
Patrizia	Olmi	Fondazione IRCCS, Istituto Nazionale dei Tumori, Milan	Italy
Ricardo	Capocaccia	Istituto Superiore di Sanità, Rome	Italy
Stefano	Ferretti	Ferrara Cancer Registry	Italy
Andrés	Cervantes	University of Valencia	Spain
Camren	Martinez-Garcia	Granja Andaluza de Salud Pública (IASPY)Granada Cancer Registry	Spain
Juan Antonio	Vizuela-Echeburu	Hospital Macarena	Spain



Vzácné nádory podle RARECARENet

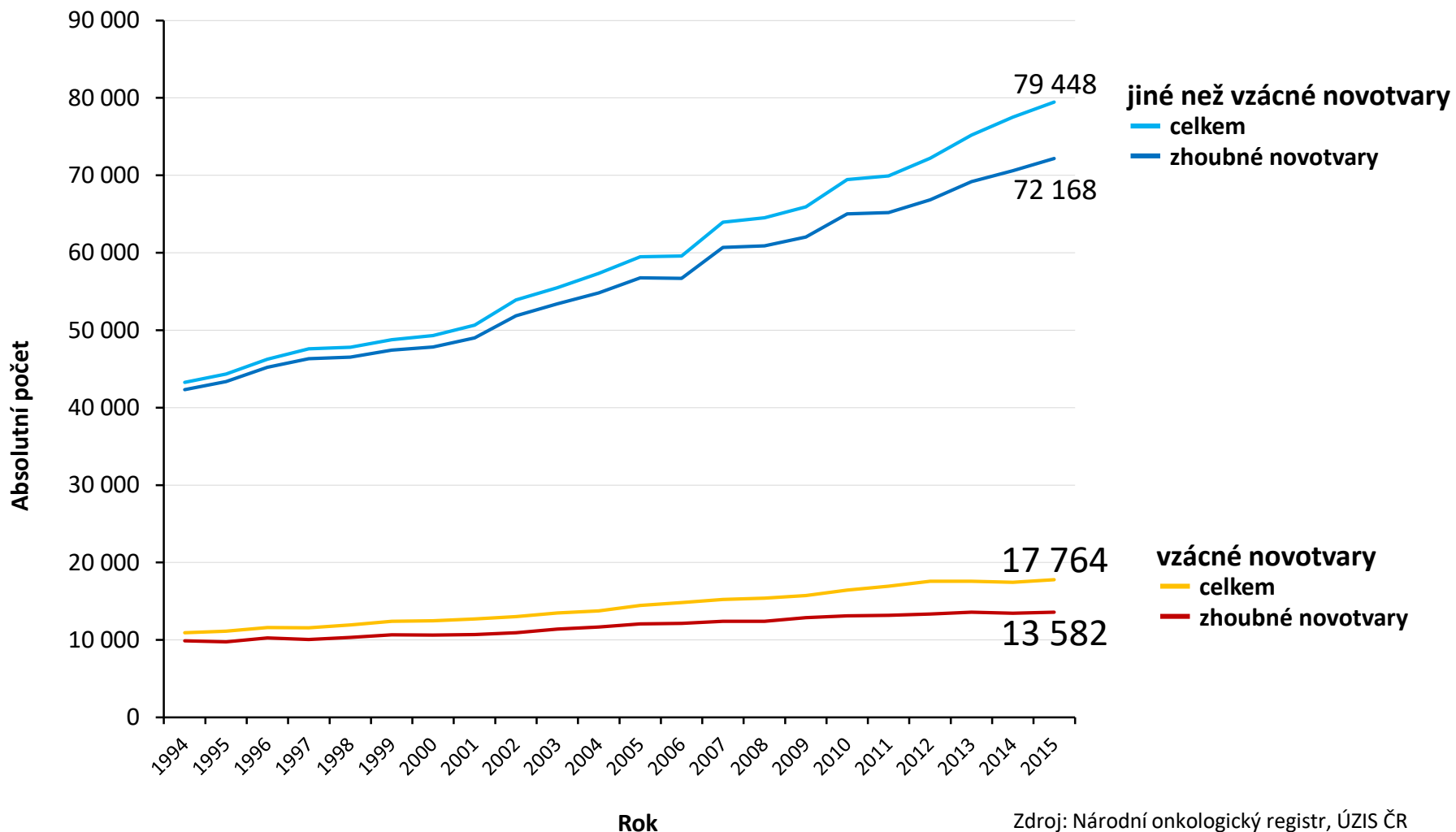
Seznam zhoubných nádorů ve 3-vrstvé struktuře
Vzácné nádory definovány pouze ve vrstvě 1 a 2

46 skupin (z 65) ve vrstvě 1
198 skupin (z 218) ve vrstvě 2

R=rare	Tier	Tumour	Crude incidence rate per 100,000	95% confidence interval		Number of cases collected in the RARECARENet database from 2000-2007	ICD-O-3 Topography code	ICD-O-3 Morphology code
								
R	1	EPITHELIAL TUMOURS OF NASAL CAVITY AND SINUSES	0,45	0,44	0,46	7 046	C30.0, C31	8000, 8001, 8003, 8004, 8010, 8011, 8012, 8020-8022, 8030, 8031, 8032, 8033-8034, 8046, 8050-8076, 8078, 8082-8084, 8120, 8121, 8122, 8123, 8130, 8140, 8144, 8560, 8980
R	2	Squamous cell carcinoma with variants of nasal cavity and sinuses	0,35	0,34	0,36	5 465	C30.0, C31	8004, 8120, 8121, 8122, 8130, 8032, 8050, 8076, 8078, 8082, 8084, 8123, 8560, 8980
	3	Squamous carcinoma					C30.0, C31	8070
	3	Verrucous carcinoma					C30.0, C31	8051
	3	Squamous cell carcinoma spindle cell					C30.0, C31	8004, 8032, 8074, 8980
	3	Papillary squamous cell carcinoma					C30.0, C31	8052
	3	Adenosquamous carcinoma					C30.0, C31	8560
	3	Squamous cell carcinoma, adenoid					C30.0, C31	8075
	3	Basaloid squamous cell carcinoma					C30.0, C31	8083
R	2	Lymphoepithelial carcinoma of nasal cavity and sinuses	0,002	0,001	0,003	31	C30.0, C31	8082
R	2	Undifferentiated carcinoma of nasal cavity and sinuses	0,02	0,02	0,02	286	C30.0, C31	8020-8022
R	2	Intestinal type adenocarcinoma of nasal cavity and sinuses	0,003	0,002	0,004	42	C30.0, C31	8144, 8140
R	1	EPITHELIAL TUMOURS OF NASOPHARYNX	0,14	0,13	0,14	2 133	C11	8000-8001, 8004, 8010-8011, 8012, 8020-8022, 8032, 8033, 8034, 8046, 8120, 8121, 8122, 8050-8076, 8078, 8082-8084, 8123, 8260, 8560, 8980
R	2	Squamous cell carcinoma with variants of nasopharynx	0,11	0,11	0,12	1 786	C11	8004, 8020-8022, 8032, 8051-8076, 8078, 8082-8084, 8120, 8121, 8122, 8123, 8560, 8980
	3	Squamous carcinoma					C11	8070
	3	Squamous cell carcinoma nonkeratinizing, NOS					C11	8072
	3	Squamous cell carcinoma keratinizing, NOS					C11	8071
	3	Papillary squamous cell carcinoma					C11	8052
	3	Basaloid squamous cell carcinoma					C11	8063
	3	Squamous cell carcinoma, adenoid					C11	8075
	3	Lymphoepithelial carcinoma					C11	8082
	3	Undifferentiated carcinoma					C11	8020-8022
R	2	Papillary adenocarcinoma of nasopharynx	0,0004	0,0001	0,001	6	C11	8050, 8260
R	1	EPITHELIAL TUMOURS OF MAJOR SALIVARY GLANDS AND SALIVARY-GI AND TYPE TUMOURS	1,39	1,37	1,41	21 794	C00-C14, C30.0, C31, C32	C00-C06, C09-C14, C30.0, C31, C32: 8140, 8141, 8143, 8144 (not in C30.0, C31), 8147, 8190, 8200, 8201, 8211, 8230, 8231, 8261, 8263, 8320, 8323, 8401, 8490, 8510, 8551, 8570, 8574, 8675, 8290, 8310, 8430, 8440, 8450, 8480, 8481, 8500, 8535, 8550, 8562, 8941, 8982,

Rare cancers are those with an incidence < 6/100,000
rare cancers are 198 (considering tier 2 only)

Vývoj incidence novotvarů dle NOR



Zdroj: Národní onkologický registr, ÚZIS ČR

Národní onkologický registr

populační registr záznamů jednotlivých novotvarů je provozován ÚZIS ČR od roku 1976

Klasifikace: MKN-10, **MKN-O-3.2 (topografie, morfologie)**, TNM

- zhoubné novotvary (C00–C97),
- novotvary in situ (D00–D09),
- novotvary nejistého nebo neznámého chování (D37–D48),

vybrané nezhoubné / benigní novotvary s morfologií *8683/0 gangliocytický paragangliom* nebo *8936/0 gastrointestinální stromální tumor, benigní* (diagnózy D10.5–D13.3, D13.5–D13.6, D13.9–D15.9, D20.0–D21.9, D28.2–D28.7, D29.1–D29.2, D30.0–D30.4, D30.9, D32.0–D35.9, D36.7 s morfologií 8683/0 nebo 8936/0).

a dále pozitivní bioptické či cytologické nálezy:

- D12 - nezhoubný novotvar tlustého střeva, konečníku, řitního kanálu a řiti,
- N87 - dysplazie hrdla děložního.

Děkuji za pozornost.

