

Seltene Tumorerkrankungen (Rare Cancer)

Häufigkeiten und Inzidenzen im Einzugsgebiet des Tumorregisters München (TRM)

RARECARENet definiert seltene Tumoren als diejenigen mit einer Inzidenz $< 6/100.000/\text{Jahr}$. Laut RARECARENet entfallen ca. 22% der Gesamtinzidenz auf diese seltenen Tumoren.

<http://www.rarecarenet.eu/>

<http://www.rarecarenet.eu/rarecarenet/index.php/cancerlist>

--> siehe List of Rare Cancers: RARECAREnet_list_of_rare_cancers_Dec15.xlsx

EUROCARE ist ein internationales kooperatives Projekt für Überlebenszeitanalysen in Europa (<http://www.eurocare.it/Home/tabid/36/Default.aspx>). Die fünfte und aktuelle Ausgabe, EUROCARE-5, umfasst Daten über mehr als 21 Millionen Tumordiagnosen, zur Verfügung gestellt von 116 Krebsregistern aus 30 europäischen Ländern.

Das **RARECARENet-Projekt** nutzt diese Datensammlung des EUROCARE-5-Projekts von 94 Krebsregistern, welche sich an EUROCARE-5 beteiligt, die RARECARENet-Qualitätskriterien erfüllt und einer Datennutzung für das RARECARENet-Projekt zugestimmt haben, Diagnosedatum 2000 bis 2007. Das Tumorregister München (**TRM**) hat sich an EUROCARE-5 und am RARECARENet-Projekt beteiligt.

Dargestellt sind im Folgenden für die von RARECARENet definierten 198 seltenen Tumorerkrankungen, die im TRM erfasste Anzahl der Diagnosen aus den Jahren 2007 bis 2021, die daraus gemittelte rohe Inzidenz pro Jahr und zum Vergleich die von RARECARENet publizierte jährliche rohe Inzidenz, gemittelt aus den Jahren 2000 bis 2007, gemeldet von 83 Krebsregistern.

Dabei entsprechen 78.742 von 322.332 Diagnosen des TRM aus den Jahren 2007 bis 2021 den von RARECARENet definierten Entitäten (24.4%)

Nr	Tumor-Entität	Anzahl n=78742	TRM Rohe Inz.	Rare Care Inz.
1	Squamous cell carcinoma with variants of nasal cavity and sinuses	322	0.433	0.3500
2	Lymphoepithelial carcinoma of nasal cavity and sinuses	0		0.0020
3	Undifferentiated carcinoma of nasal cavity and sinuses	13	0.017	0.0200
4	Intestinal type adenocarcinoma of nasal cavity and sinuses	49	0.066	0.0030
5	Squamous cell carcinoma with variants of nasopharynx	190	0.256	0.1100
6	Papillary adenocarcinoma of nasopharynx	0		0.0004
7	Epithelial tumours of major salivary glands	660	0.888	0.9600
8	Salivary gland type tumours of head and neck	192	0.258	0.4300
9	Squamous cell carcinoma with variants of hypopharynx	1112	1.496	1.2700
10	Squamous cell carcinoma with variants of larynx	1667	2.243	4.6100
11	Squamous cell carcinoma with variants of oropharynx	2204	2.966	3.1200
12	Squamous cell carcinoma with variants of oral cavity	2162	2.909	3.5100
13	Squamous cell carcinoma with variants of lip	244	0.328	1.0200
14	Squamous cell carcinoma with variants of oesophagus	2215	2.981	3.3600
15	Adenocarcinoma with variants of oesophagus	1384	1.862	3.2600
16	Salivary gland type tumours of oesophagus	0		0.0040
17	Undifferentiated carcinoma of oesophagus	20	0.027	0.0400
18	Squamous cell carcinoma with variants of stomach	44	0.059	0.1200
19	Salivary gland-type tumours of stomach	0		0.0020
20	Undifferentiated carcinoma of stomach	45	0.061	0.2100
21	Adenocarcinoma with variants of small intestine	481	0.647	0.5900
22	Squamous cell carcinoma with variants of small intestine	1	0.001	0.0100
23	Squamous cell carcinoma with variants of colon	11	0.015	0.0300
24	Fibromixoma and low grade mucinous adenocarcinoma (pseudomixoma peritonei) of the appendix	111	0.149	0.1100
25	Squamous cell carcinoma with variants of rectum	29	0.039	0.1100
26	Squamous cell carcinoma with variants of anal canal	1302	1.752	0.8100
27	Adenocarcinoma with variants of anal canal	66	0.089	0.2500
28	Paget's disease of anal canal	0		0.0010
29	Squamous cell carcinoma with variants of pancreas	28	0.038	0.0200
30	Acinar cell carcinoma of pancreas	53	0.071	0.0300
31	Mucinous cystadenocarcinoma of pancreas	41	0.055	0.0100
32	Intraductal papillary mucinous carcinoma invasive of pancreas	58	0.078	0.0100
33	Solid pseudopapillary carcinoma of pancreas	14	0.019	0.0030
34	Serous cystadenocarcinoma of pancreas	4	0.005	0.0003
35	Carcinoma with osteoclast-like giant cells of pancreas	1	0.001	0.0010
36	Hepatocellular carcinoma of Liver and IBT	3642	4.901	3.2200
37	Hepatocellular carcinoma, fibrolamellar	15	0.020	0.0200

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38	Cholangiocarcinoma of IBT	997	1.342	0.9700
39	Adenocarcinoma with variants of liver and IBT	283	0.381	0.4100
40	Undifferentiated carcinoma of liver and IBT	8	0.011	0.0200
41	Squamous cell carcinoma with variants of liver and IBT	2	0.003	0.0100
42	Bile duct cystadenocarcinoma of IBT	0		0.0020
43	Adenocarcinoma with variants of gallbladder	802	1.079	1.3500
44	Adenocarcinoma with variants of EBT	1862	2.506	1.4400
45	Squamous cell carcinoma of gallbladder and EBT	15	0.020	0.0300
46	Squamous cell carcinoma with variants of trachea	28	0.038	0.0600
47	Adenocarcinoma with variants of trachea	7	0.009	0.0100
48	Salivary gland type tumours of trachea	9	0.012	0.0100
49	Adenosquamous carcinoma of lung	279	0.375	0.2900
50	Large cell carcinoma of lung	329	0.443	3.8100
51	Salivary gland type tumours of lung	39	0.052	0.0600
52	Sarcomatoid carcinoma of lung	190	0.256	0.2100
53	Malignant thymoma	177	0.238	0.1400
54	Squamous cell carcinoma of thymus	47	0.063	0.0100
55	Undifferentiated carcinoma of thymus	4	0.005	0.0020
56	Lymphoepithelial carcinoma of thymus	0		0.0010
57	Adenocarcinoma with variants of thymus	6	0.008	0.0030
58	Mammary Paget's disease of breast	59	0.079	0.4100
59	Special types of adenocarcinoma of breast	1877	2.526	3.0600
60	Metaplastic carcinoma of breast	179	0.241	0.1000
61	Salivary gland type tumours of breast	47	0.063	0.0600
62	Epithelial tumour of male breast	443	1.205	1.0600
63	Squamous cell carcinoma with variants of corpus uteri	8	0.011	0.0600
64	Adenoid cystic carcinoma of corpus uteri	0		0.0003
65	Clear cell adenocarcinoma, NOS	193	0.260	0.1600
66	Serous (papillary) carcinoma	276	0.371	0.0800
67	Mullerian mixed tumour	307	0.413	0.4000
68	Squamous cell carcinoma with variants of cervix uteri	2412	3.246	4.7300
69	Adenocarcinoma with variants of cervix uteri	654	0.880	0.9100
70	Undifferentiated carcinoma of cervix uteri	8	0.011	0.0300
71	Mullerian mixed tumour of cervix uteri	8	0.011	0.0200
72	Adenocarcinoma with variants of ovary	3976	5.351	5.9500
73	Mucinous adenocarcinoma of ovary	289	0.389	0.7700
74	Clear cell adenocarcinoma of ovary	218	0.293	0.3000
75	Primary peritoneal serous/papillary carcinoma	476	0.641	0.0800
76	Mullerian mixed tumour of ovary	77	0.104	0.1400
77	Adenocarcinoma with variants of fallopian tube	205	0.276	0.1700
78	Sex cord tumours of ovary	134	0.180	0.1300
79	Malignant/Immature teratomas of ovary	20	0.027	0.0500
80	Germ cell tumour of ovary	57	0.077	0.0700
81	Squamous cell carcinoma with variants of vulva and vagina	1637	2.203	1.6900
82	Adenocarcinoma with variants of vulva and vagina	45	0.061	0.0700
83	Paget's disease of vulva and vagina	22	0.030	0.0500
84	Undifferentiated carcinoma of vulva and vagina	2	0.003	0.0100
85	Choriocarcinoma of placenta	14	0.019	0.0200

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86	Squamous cell carcinoma with variants of prostate	9	0.012	0.0200
87	Infiltrating duct carcinoma of prostate	678	0.912	0.5100
88	Transitional cell carcinoma of prostate	11	0.015	0.0600
89	Basal cell adenocarcinoma of prostate	2	0.003	0.0100
90	Paratesticular adenocarcinoma with variants	0		0.0010
91	Non seminomatous testicular cancer	678	0.912	1.2700
92	Seminomatous testicular cancer	2015	2.712	1.8200
93	Spermatocytic seminoma	12	0.016	0.0300
94	Teratoma with malignant transformation	1	0.001	0.0010
95	Testicular sex cord cancer	8	0.011	0.0200
96	Squamous cell carcinoma with variants of penis	408	0.549	0.6200
97	Adenocarcinoma with variants of penis	1	0.001	0.0100
98	Squamous cell carcinoma spindle cell type of kidney	7	0.009	0.0100
99	Squamous cell carcinoma with variants of kidney	15	0.020	0.0300
100	Transitional cell carcinoma of pelvis and ureter	1543	2.076	1.4100
101	Squamous cell carcinoma with variants of pelvis and ureter	30	0.040	0.0200
102	Adenocarcinoma with variants of pelvis and ureter	8	0.011	0.0200
103	Transitional cell carcinoma of urethra	162	0.218	0.0900
104	Squamous cell carcinoma with variants of urethra	27	0.036	0.0200
105	Adenocarcinoma with variants of urethra	6	0.008	0.0100
106	Squamous cell carcinoma with variants of bladder	246	0.331	0.3600
107	Adenocarcinoma with variants of bladder	104	0.140	0.3000
108	Salivary gland type tumours of bladder	1	0.001	0.0000
109	Squamous cell carcinoma with variants of eye and adnexa	32	0.043	0.0300
110	Adenocarcinoma with variants of eye and adnexa	6	0.008	0.0100
111	Squamous cell carcinoma with variants middle ear	4	0.005	0.0200
112	Adenocarcinoma with variants of middle ear	2	0.003	0.0030
113	Mesothelioma of pleura and pericardium	1102	1.483	1.8300
114	Mesothelioma of peritoneum and tunica vaginalis	104	0.140	0.1300
115	Malignant melanoma of mucosa and extracutaneous	173	0.233	0.1500
116	Malignant melanoma of uvea	297	0.400	0.7000
117	Adnexal carcinoma of skin	2	0.003	0.3000
118	Neuroblastoma and ganglioneuroblastoma	38	0.051	0.1000
119	Nephroblastoma	82	0.110	0.1300
120	Retinoblastoma	13	0.017	0.0500
121	Heptatoblastoma	19	0.026	0.0200
122	Pleuropulmonary blastoma	1	0.001	0.0010
123	Pancreatoblastoma	0		0.0020
124	Olfactory neuroblastoma	27	0.036	0.0300
125	Odontogenic malignant tumours	2	0.003	0.0040
126	Non seminomatous germ cell tumours	40	0.054	0.0600
127	Seminomatous germ cell tumors	20	0.027	0.0100
128	Germ cell tumors of Central Nervous System (CNS)	18	0.024	0.0400
129	Soft tissue sarcoma of head and neck	226	0.304	0.2600
130	Soft tissue sarcoma of limbs	885	1.191	1.1000
131	Soft tissue sarcoma of superficial trunk	392	0.528	0.5000
132	Soft tissue sarcoma of mediastinum	17	0.023	0.0300
133	Soft tissue sarcoma of heart	18	0.024	0.0100

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134	Soft tissue sarcoma of breast	108	0.145	0.1800
135	Soft tissue sarcoma of uterus	381	0.513	0.5500
136	Soft tissue sarcoma of paratestis	35	0.047	0.0300
137	Soft tissue sarcomas of other genitourinary tract (vulva, vagina, ovary, penis, prostate, testis, kidney, renal pelvis, ureter, bladder, urethra)	116	0.156	0.2000
138	Soft tissue sarcoma of viscera	237	0.319	0.3800
139	Soft tissue sarcoma of retroperitoneum and peritoneum	256	0.345	0.3100
140	Soft tissue sarcoma of pelvis	182	0.245	0.2000
141	Soft tissue sarcoma of skin	3	0.004	0.3000
142	Soft tissue sarcoma of paraorbit	3	0.004	0.0100
143	Soft tissue sarcoma of brain and other parts of the nervous system	142	0.191	0.1700
144	Embryonal rhabdomyosarcoma of soft tissue	25	0.034	0.0500
145	Alveolar rhabdomyosarcoma of soft tissue	21	0.028	0.0300
146	Ewing's sarcoma of soft tissue	33	0.044	0.0600
147	Osteogenic sarcoma	137	0.184	0.2100
148	Chondrogenic sarcomas	169	0.227	0.2600
149	Notochordal sarcomas, chordoma	46	0.062	0.0700
150	Vascular sarcomas	2	0.003	0.0100
151	Ewing's sarcoma	75	0.101	0.1200
152	Epithelial tumours, adamantinoma	2	0.003	0.0100
153	Other high grade sarcomas (fibrosarcoma, malignant fibrous histiocytoma)	4	0.005	0.0200
154	Gastrointestinal stromal sarcoma	1451	1.953	0.3000
155	Kaposi's sarcoma	61	0.082	0.2500
156	Well differentiated not functioning endocrine carcinoma of pancreas and digestive tract	7	0.009	1.0100
157	Well differentiated functioning endocrine carcinoma of pancreas and digestive tract	0		0.0300
158	Poorly differentiated endocrine carcinoma of pancreas and digestive tract	4	0.005	0.6700
159	Mixed endocrine-exocrine carcinoma of pancreas and digestive tract	0		0.0100
160	Endocrine carcinoma of thyroid gland	312	0.420	0.2400
161	Neuroendocrine carcinoma of skin	2	0.003	0.1900
162	Typical and atypical carcinoid of the lung	619	0.833	0.3900
163	Neuroendocrine carcinoma of other sites	960	1.292	0.9000
164	Pheochromocytoma, malignant	26	0.035	0.0400
165	Paraganglioma	19	0.026	0.0200
166	Carcinoma of pituitary gland	33	0.044	0.0400
167	Carcinoma of thyroid gland	6280	8.451	5.0700
168	Carcinomas of parathyroid gland	16	0.022	0.0300
169	Carcinoma of adrenal cortex	138	0.186	0.2200
170	Astrocytic tumours of CNS	3347	4.504	4.9900
171	Oligodendroglial tumours of CNS	124	0.167	0.3900
172	Ependymal tumours of CNS	132	0.178	0.2100
173	Neuronal and mixed neuronal-glial tumors	91	0.122	0.0050
174	Choroid plexus carcinoma of CNS	2	0.003	0.0100
175	Malignant meningiomas	57	0.077	0.1600
176	Embryonal tumors of CNS	6	0.008	0.2000

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177	Hodgkin lymphoma, classical	1355	1.823	2.4600
178	Hodgkin lymphoma nodular lymphocyte predominance	109	0.147	0.0900
179	Precursor B/T lymphoblastic leukaemia/lymphoblastic lymphoma (and Burkitt leukemia/lymphoma)	800	1.077	1.4600
180	T cutaneous lymphoma (Sezary syn, Mycosis fung)	114	0.153	0.3500
181	Other T cell lymphomas and NK cell neoplasms	574	0.772	0.6200
182	Diffuse B lymphoma	3582	4.820	4.3200
183	Follicular B lymphoma	1732	2.331	2.1900
184	Hairy cell leukaemia	167	0.225	0.2800
185	Plasmacytoma/Multiple Myeloma (and Heavy chain diseases)	3554	4.783	5.7100
186	Mantle cell lymphoma	590	0.794	0.5600
187	Prolymphocytic leukaemia, B cell	6	0.008	0.0500
188	Acute promyelocytic leukemia (AML with t(15,17) with variants	51	0.069	0.1200
189	AML	2707	3.643	3.5000
190	Chronic myeloid leukemia	798	1.074	1.1200
191	Other myeloproliferative neoplasms	1	0.001	2.1700
192	Mast cell tumour	32	0.043	0.0300
193	Myelodysplastic syndrome with 5q syndrome	0		0.0100
194	Other myelodysplastic syndrome	0		2.1400
195	Chronic Myelomonocytic leukemia	238	0.320	0.2900
196	Atypical chronic myeloid leukemia BCR/ABL negative	9	0.012	0.0200
197	Histiocytic malignancies	51	0.069	0.0400
198	Lymph node accessory cell tumors	9	0.012	0.0100