R A R E C A N C E R

始まった
希少がん対策

2017年11月25日(土)
13:00 – 17:00
日本医師会館大講堂

「欧州での希少がん対策
—RARECAREの経験より」

アナリザ・トラマ
分析疫学ユニット
ヘルスケア科学研究所　国立がん研究所　（ミラノ、イタリア）

Sistema Socio Sanitario
IRCCS Istituto Nazionale dei Tumori
Regione Lombardia

JARC
JOINT ACTION ON RARE CANCERS

本講演のねらい

1. ヨーロッパで提唱された希少がん定義と希少がんリスト

2. 日本における希少がんの負担の説明と、がん登録の国際標準化・共同調査の重要性

3. ヨーロッパでの希少がんの扱いに関する情報提供

希少がん定義

目的

RARECARE
Surveillance of Rare Cancers in Europe

- 「希少がん」の定義と希少がんリスト
- ヨーロッパでの希少がんの負担を推計する
- がん登録のデータの精度を向上させる
- キーパーソンに対して情報を周知するための方法を画策する

期間：3年間（開始日2007年4月1日）

希少がん

罹患<6/100,000/年（EU）

EUROPEAN JOURNAL OF CANCER 47 (2011) 2493–2511

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ELSEVIER

EJC

Rare cancers are not so rare: The rare cancer burden in Europe

Gemma Gatta [a,], Jan Maarten van der Zwan [b], Paolo G. Casali [c], Sabine Siesling [b], Angelo Paolo Dei Tos [d], Ian Kunkler [e], Renée Otter [b], Lisa Licitra [f], Sandra Mallone [g], Andrea Tavilla [g], Annalisa Trama [a], Riccardo Capocaccia [g], The RARECARE working group*

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希少疾患：
有病数<50/100,000/年（EU）

動機

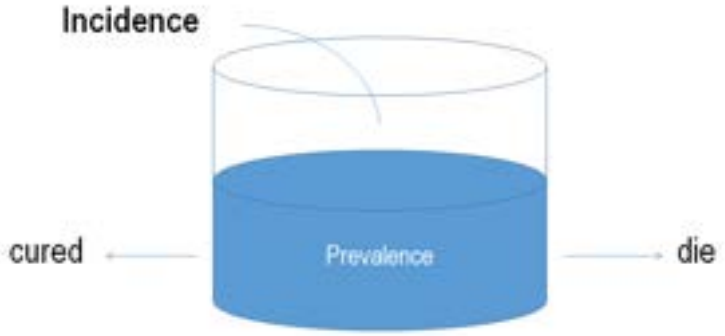
RARECARE
Surveillance of Rare Cancers in Europe

頻度

- 希少がんは、臨床判断、医療管理・政策、臨床研究において、その発生数の少なさから問題を抱えているがんである

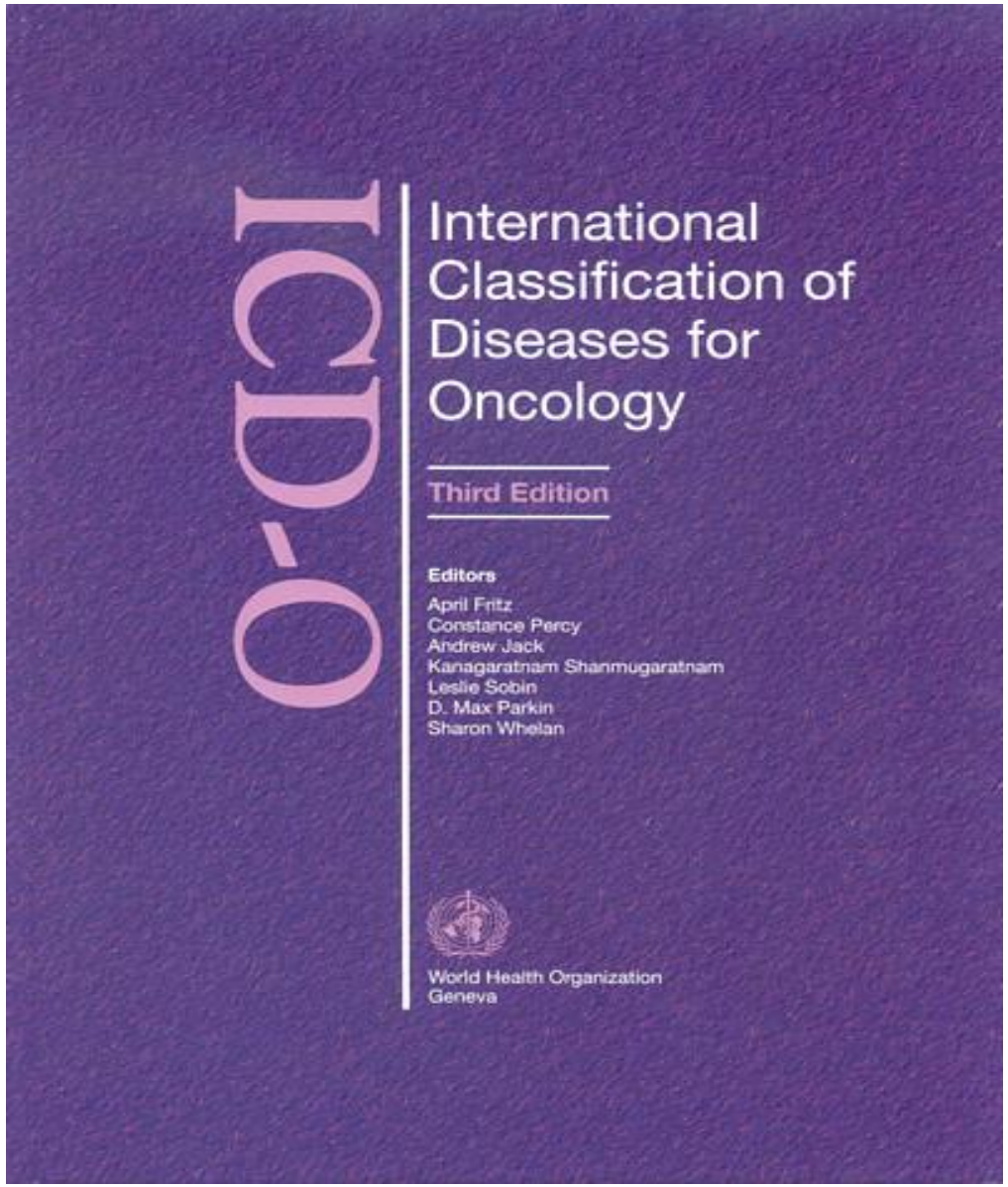
罹患

- 希少がんには罹患が指標として適している
- がん以外の希少疾患には、有病数の方が指標として適している
- 何を最適な指標とするかは治療に関連している
- オーファンドラッグ（希少疾病用医薬品）の認可への関心



希少がんリスト

RARECARE
Surveillance of Rare Cancers in Europe



- 新しい分類ではない
- ICD-Oの局在と形態コードに準拠
- 階層構造

階層構造

RARECARE
Surveillance of Rare Cancers in Europe

- 第3層⇒全てのWHO分類の腫瘍
- 第2層⇒臨床家にとって単独の疾患として捉えられるもの
  - 臨床的判断；臨床研究
- 第1層⇒同じようなクリニカルパスをたどる腫瘍
  - 医療管理・政策

希少がんリストの構成

RARECARE
Surveillance of Rare Cancers in Europe

R=rare	Tier	Tumour	Topography	Morphology code
	1	TESTICULAR AND PARATESTICULAR CANCERS	C62, C63.0, C63.1, C63.8	8000, 8001,8010,8011, 8120, 8123, 8140-8141, 8147, 8190, 8210-8211, 8221, 8231, 8255, 8260, 8261-8263,
R	2	**Paratesticular adenocarcinoma with variants**	C63.0, C63.1	8120, 8123, 8140-8141, 8147, 8190, 8200, 8210-8211,
	3	Endometrioid adenocarcinoma, NOS	C63.0, C63.1	8380
	3	Clear cell adenocarcinoma, NOS	C63.0, C63.1	8310
	3	Serous cy		
	3	Mucinous		
	3	Collecting		orphology code
	3	Transitiona		
R	2	**Non seminom**		, 9065, 9070-9072
	3	Mixed ger		
	3	Teratocarc		
	3	Choriocarc		
	3	Embryona		
	3	Yolk sac t		
	3	Choriocarc		
R	2	**Seminomatou**		
	3	Seminoma		
R	2	**Spermatocyti**		
R	2	**Teratoma with malignant transformation**		
R	2	**Testicular sex cord cancer**	C62	8630-8640, 8650, 8590-8592
	1	EPITHELIAL TUMOURS OF PENIS	C60	8000-8001, 8010-8011, 8020-8022, 8050-8084, 8090,
R	2	**Squamous cell carcinoma with variants of penis**	C60	8020-8022, 8050-8084, 8123
	3	Squamous carcinoma	C60	8070
	3	Verrocous carcinoma	C60	8051
	3	Squamous cell carcinoma, sarcomatoid	C60	8074
	3	Adenosquamous carcinoma	C60	8560
	3	Basaloid carcinoma	C60	8123
R	2	**Adenocarcinoma with variants of penis**	C60	8140-8141, 8147, 8190, 8200-8201, 8210-8211, 8230-8231, 8255-8263, 8310, 8323, 8440, 8480-8490, 8504,
	3	Extramammary Paget's disease	C60	8542

198の異なる希少がん（第2層）

上皮がん（epithelial）は表面的（epithelial）ではない

Tier	Tumour	Topography code	Morphology code
1	EPITHELIAL TUMOURS OF NASAL CAVITY AND SINUSES	C30.0, C31	8000, 8001, 8004, 8010, 8011, 8020-8022, 8032,
1	EPITHELIAL TUMOURS OF OESOPHAGUS	C15	8000-8001, 8004, 8010-8011, 8020-8022, 8032,
1	EPITHELIAL TUMOUR OF TRACHEA	C33	8000-8001, 8004, 8010-8011, 8044, 8020-8022,
1	EPITHELIAL TUMOURS OF CORPUS UTERI	C54	8000, 8001, 8010, 8011, 8070-8078, 8082-8084,
1	EPITELIAL TUMOURS OF PROSTATE	C61, C637	8000-8001, 8020-8022, 8032, 8050, 8070-8072,
1	EPITELIAL TUMOURS OF KIDNEY	C64	8000-8001, 8010-8011, 8020, 8022, 8030-8035,
1	EPITHELIAL TUMOURS OF EYE AND ADNEXA	C69	8000-8001, 8010-8011, 8020, 8050-8084, 8090,
1	EMBRYONAL NEOPLASMS	all cancers sites,	8960, 8970-8973, 9490, 9500, 9510-9514
1	EXTRAGONADAL GERM CELL TUMOURS	all cancers sites	9060-9072, 9080-9085, 9101, and 9100 if not in
1	SOFT TISSUE SARCOMA	all cancers sites	8800-8935, 8940, 8950-8959, 8963-8964, 8990-
	NEUROENDOCRINE TUMOURS	all cancer sites	8013, 8041-8045, 8150-8157, 8240-8247, 8249,
1	GLIAL TUMOURS OF CENTRAL NERVOUS SYSTEM (CNS)	C71, C72.0, C72.8-	9380-9384, 9391-9460
1	NON GLIAL TUMOURS OF CNS AND PINEAL GLAND	C71, C72.0, C75.3	9362, 9390, 9470-9474, 9490, 9500-9505, 9508
1	MALIGNANT MENINGIOMAS	C70	9530, 9538-9539
1	LYMPHOID DISEASES		9590, 9591, 9596, 9650-9655, 9659, 9661-9667,
1	ACUTE MYELOID LEUKEMIA AND RELATED PRECURSOR NEOPLASMS		9805, 9840, 9860-9861, 9866-9867, 9870-9874, 9891, 9895-9897, 9910, 9920, 9930-9931, 9984,
1	MYELOPROLIFERATIVE NEOPLASMS		9740-9742, 9863, 9875, 9950, 9960-9964

リスト作成までの道のり

RARECARE
Surveillance of Rare Cancers in Europe

- RARECAREグループの専門家からの提案
  - 病理学者、腫瘍学者、疫学者
- コンセンサス・ワークショップ
  - ブリュッセルとトレヴィーゾにて
- WEBでの討論
- 学術団体の合意

コンセンサス・ワークショップ

学術団体

ヨーロッパ病理学会

ヨーロッパ外科腫瘍学会

ヨーロッパ放射線腫瘍学会

ヨーロッパ臨床腫瘍学会

ヨーロッパ小児がん学会

ヨーロッパがん治療研究会

ヨーロッパがん研究所会

ヨーロッパがん学校

ヨーロッパがん患者連合

部位別専門家

頭頸部がん

胸部がん

消化器がん

婦人科がん

泌尿器・男性生殖器がん

肉腫

神経内分泌がん

血液がん

Home
Project Aims
Work Packages
Rare Cancers
Meetings
Other Activities
Contact Us

List of Rare Cancers

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

Please download the file above and then send us an email with your comments.

Send us an email with your comments

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 1995-2002 and the corresponding incidence rates. Both figures are derived from the data of 70 population-based cancer registries adhering to the RARECARE project.

List of Rare Cancers - download PDF

Please download the file above and then send us an email with your comments.

Send us an email with your comments

希少がん

1. **頭頸部がん**（鼻腔･副鼻腔がん、**上咽頭がん**、下咽頭がん、喉頭、唾液腺、中咽頭、口腔、口唇、眼、中耳）
2. **胸部希少がん**（気管腫瘍、胸腺腫、悪性中皮腫）
3. **男性生殖器及び泌尿器希少がん**（精巣腫瘍、陰茎、腎盂、尿道、尿管、性腺外胚細胞腫瘍）
4. **女性生殖器希少がん**（外陰部及び膣の腫瘍、卵巣の非上皮性腫瘍、胎盤絨毛がん）
5. **神経内分泌腫瘍**
6. **内分泌器官の腫瘍**（甲状腺癌、副甲状腺、副腎外皮、脳下垂体）
7. **中枢神経腫瘍**（グリア系腫瘍、髄芽腫、悪性髄膜腫）
8. **肉腫**（軟部腫瘍、骨肉腫、消化管間質腫瘍）
9. **消化器系希少がん**（小腸、肛門管、胆嚢、肝外胆管腫瘍）
10. **希少皮膚がんと皮膚以外のメラノーマ**（粘膜およびぶどう膜のメラノーマ、皮膚付属器がん、カポジ肉腫）
11. **血液希少がん**（急性骨髄性白血病、骨髄増殖性の腫瘍、骨髄異形成および骨髄増殖性の腫瘍、組織球のおよび樹状細胞腫瘍）
12. **小児がん（all）**

このリストの活用法は？

- 負担の指標を作成する
- がん登録データの質の向上
- 保健医療上の特別な配慮の必要な分野の特定

EUでの希少がんの負担

希少がんの負担指標

EUROCARE4データベース

- 88ヨーロッパがん登録（EU22カ国）
- 人口カバー：1億4600万
- 診断年：1995-2002

RARECARE
Surveillance of Rare Cancers in Europe

希少がん:罹患

EUROPEAN JOURNAL OF CANCER 47 (2011) 2493–2511

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Rare cancers are not so rare: The rare cancer burden in Europe

Gemma Gatta [a,*], *Jan Maarten van der Zwan* [b], *Paolo G. Casali* [c], *Sabine Siesling* [b], *Angelo Paolo Dei Tos* [d], *Ian Kunkler* [e], *Renée Otter* [b], *Lisa Licitra* [f], *Sandra Mallone* [g], *Andrea Tavilla* [g], *Annalisa Trama* [a], *Riccardo Capocaccia* [g], *The RARECARE working group*

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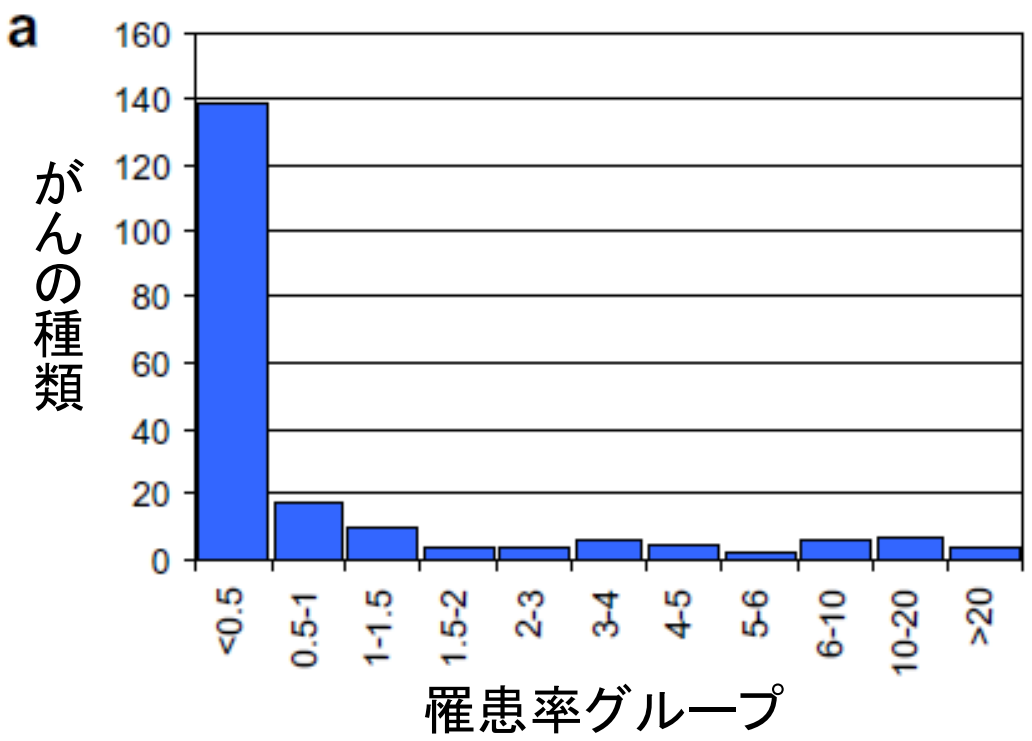
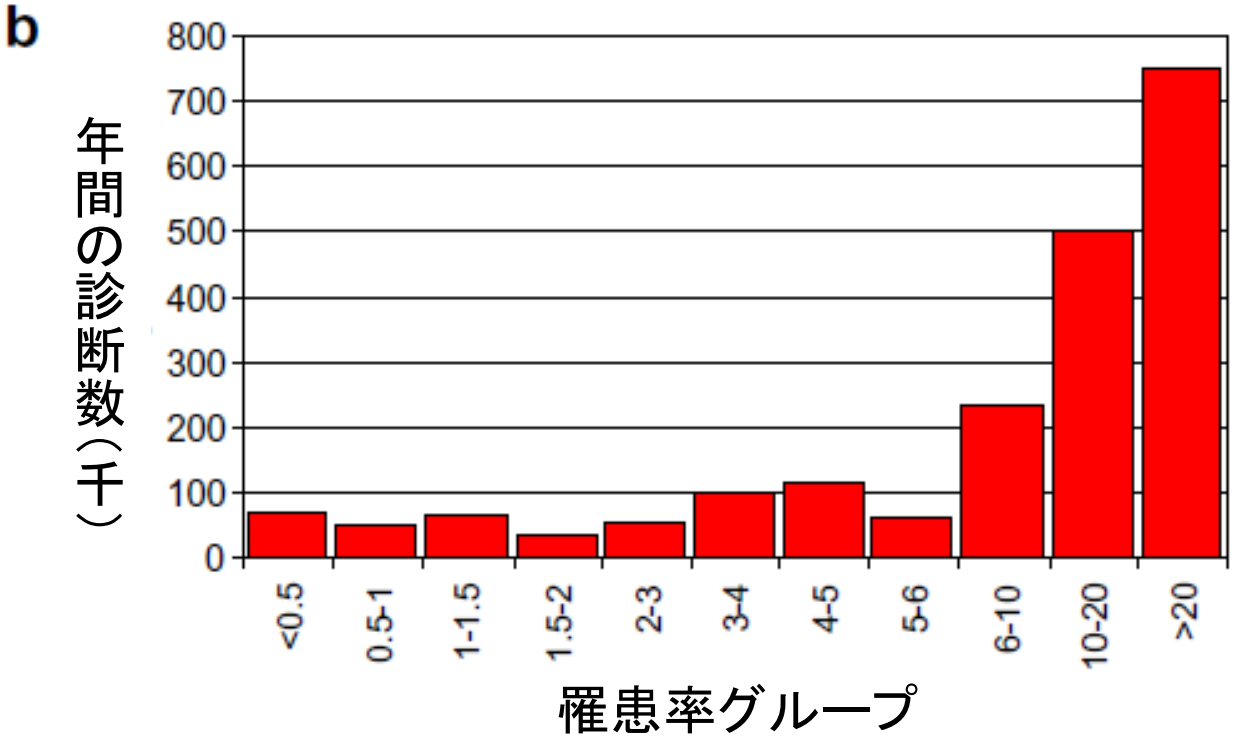
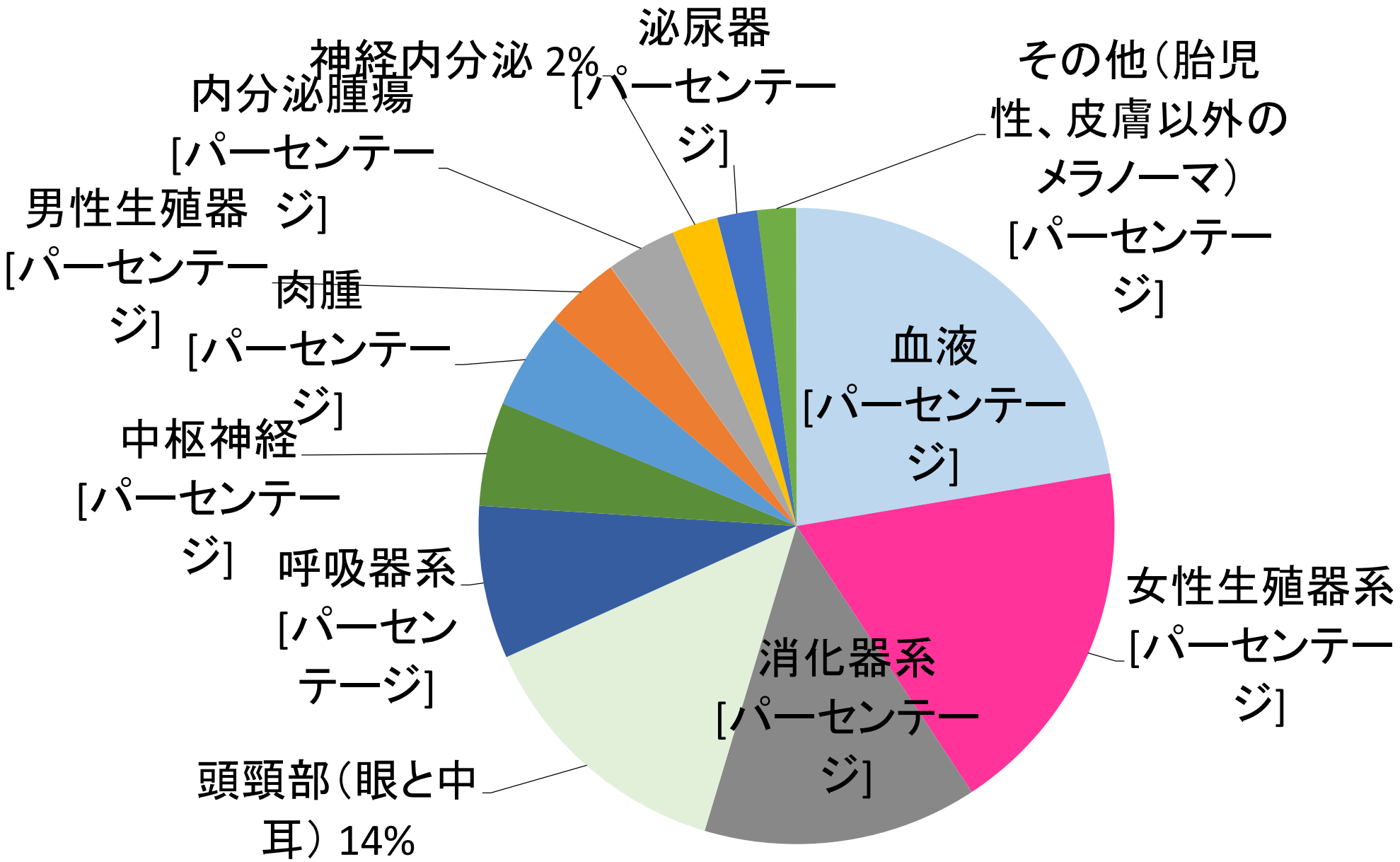
罹患率グループ別のがんの種類の分布(a)及び年間の診断数(b)EU27

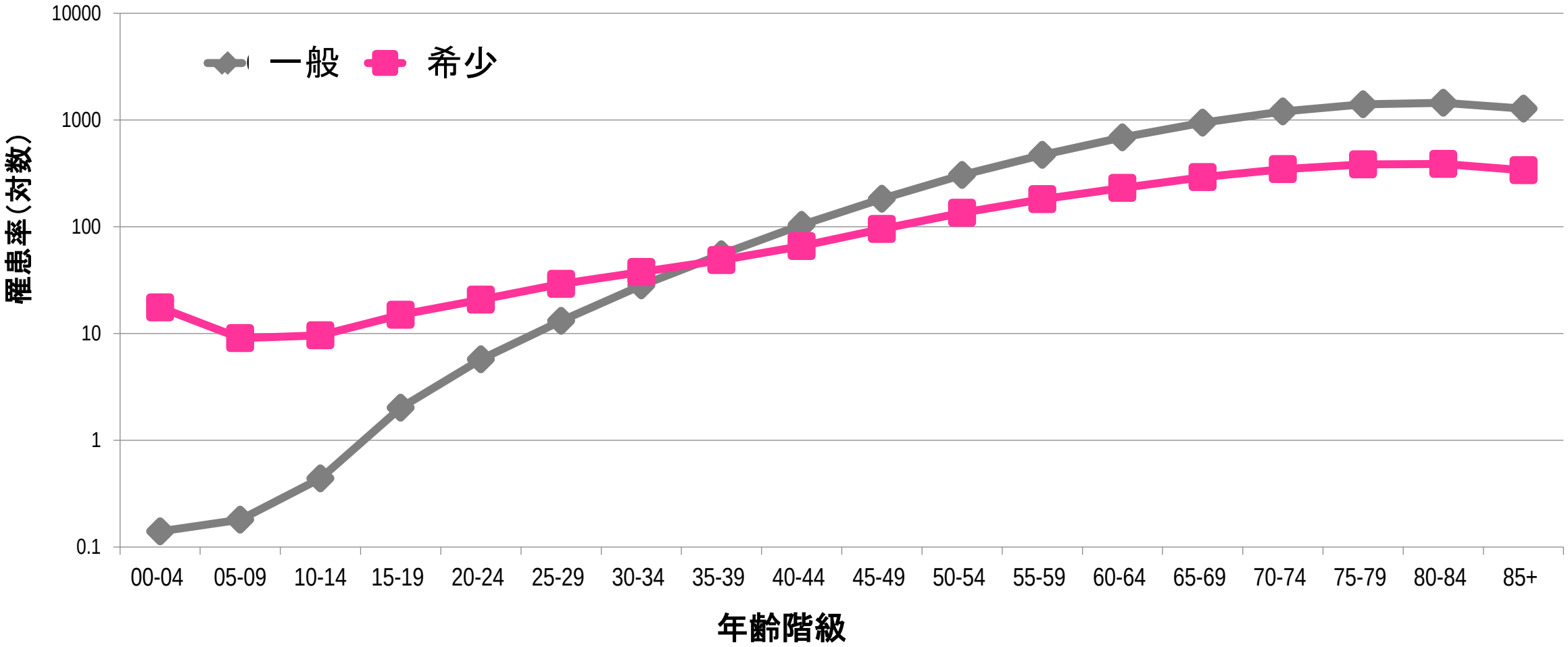




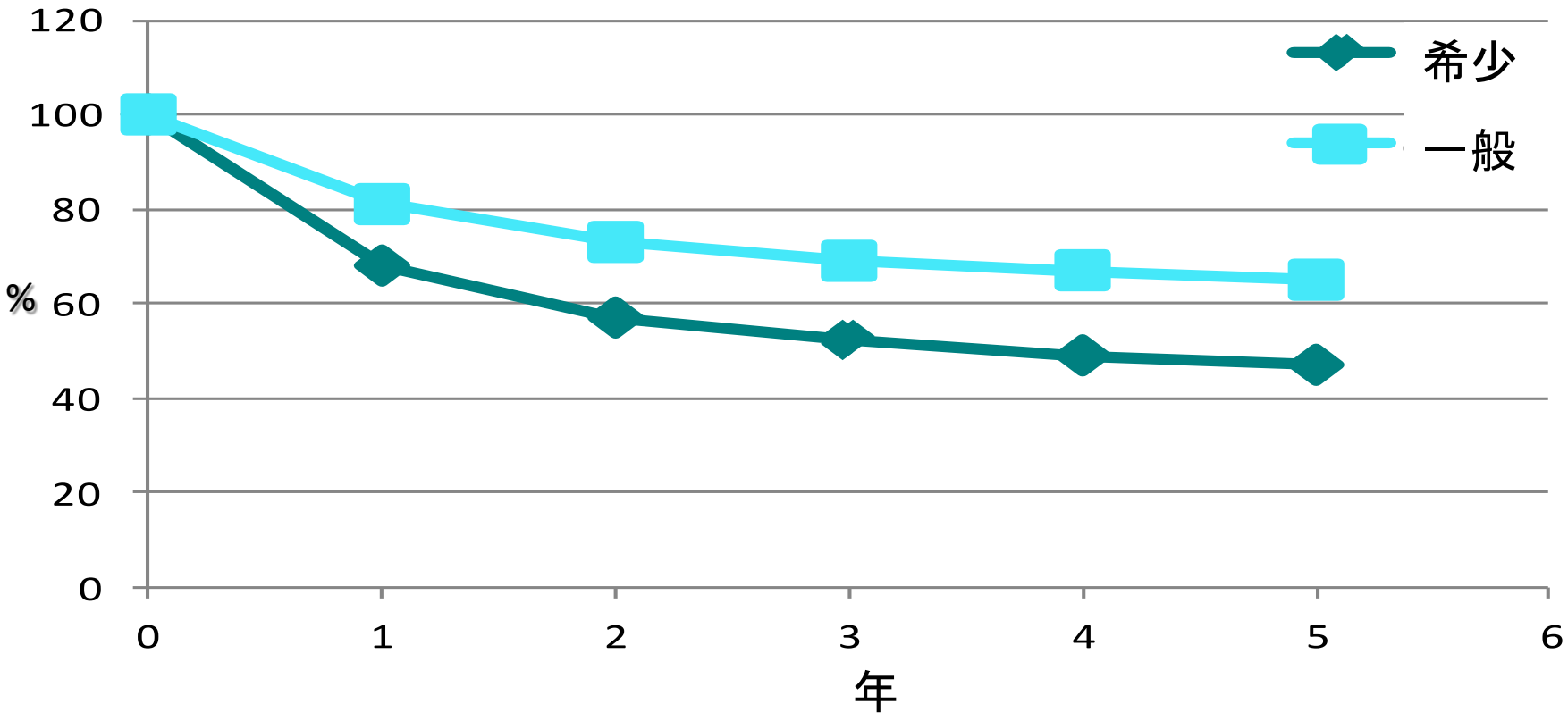
希少腫瘍の主な系統の分布（全希少がん中）



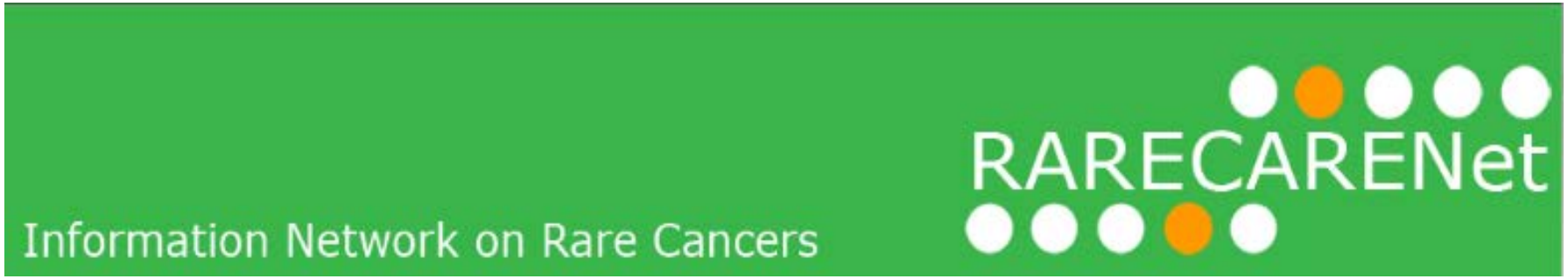
希少がんと一般的ながんの年齢調整罹患率（EU27）



希少がん:5年相対生存率




Surveillance of Rare Cancers in Europe
RARECARE


Information Network on Rare Cancers
RARECARENet

RARECAREnet（診断年2000-2007）

94がん登録（89　RARECARE）

24カ国（19　RARECARE）

ブルガリア
チェコ共和国
エストニア
ラトビア
リトアニア

東ヨーロッパ

クロアチア

カバー率：EU人口の46%（ノルウェー、スイス、アイスランド除く）

EUROCARE5　成人データベース

Burden and centralised treatment in Europe of rare tumours: results of RARECAREnet—a population-based study

*Gemma Gatta, Riccardo Capocaccia, Laura Botta, Sandra Mallone, Roberta De Angelis, Eva Ardanaz, Harry Comber, Nadya Dimitrova, Maarit K Leinonen, Sabine Siesling, Jan M van der Zwan, Liesbet Van Eycken, Otto Visser, Maja P Žakelj, Lesley A Anderson, Francesca Bella, Kaire Innos, Renée Otter, Charles A Stiller, Annalisa Trama, for the RARECAREnet working group**

Lancet Oncol 2017; 18: 1022–39

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This online publication has been corrected. The corrected version first appeared at thelancet.com/oncology on July 26, 2017

See Comment page 983

*Members of the RARECARENet Working Group are listed in the appendix

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Summary

Background Rare cancers pose challenges for diagnosis, treatments, and clinical decision making. Information about rare cancers is scant. The RARECARE project defined rare cancers as those with an annual incidence of less than six per 100 000 people in European Union (EU). We updated the estimates of the burden of rare cancers in Europe, their time trends in incidence and survival, and provide information about centralisation of treatments in seven European countries.

Methods We analysed data from 94 cancer registries for more than 2 million rare cancer diagnoses, to estimate European incidence and survival in 2000–07 and the corresponding time trends during 1995–2007. Incidence was calculated as the number of new cases divided by the corresponding total person-years in the population. 5-year relative survival was calculated by the Ederer-2 method. Seven registries (Belgium, Bulgaria, Finland, Ireland, the Netherlands, Slovenia, and the Navarra region in Spain) provided additional data for hospitals treating about 220 000 cases diagnosed in 2000–07. We also calculated hospital volume admission as the number of treatments provided by each hospital rare cancer group sharing the same referral pattern.

Findings Rare cancers accounted for 24% of all cancers diagnosed in the EU during 2000–07. The overall incidence rose annually by 0·5% (99·8% CI 0·3–0·8). 5-year relative survival for all rare cancers was 48·5% (95% CI 48·4 to 48·6), compared with 63·4% (95% CI 63·3 to 63·4) for all common cancers. 5-year relative survival increased (overall 2·9%, 95% CI 2·7 to 3·2), from 1999–2001 to 2007–09, and for most rare cancers, with the largest increases for haematological tumours and sarcomas. The amount of centralisation of rare cancer treatment varied widely between cancers and between countries. The Netherlands and Slovenia had the highest treatment volumes.

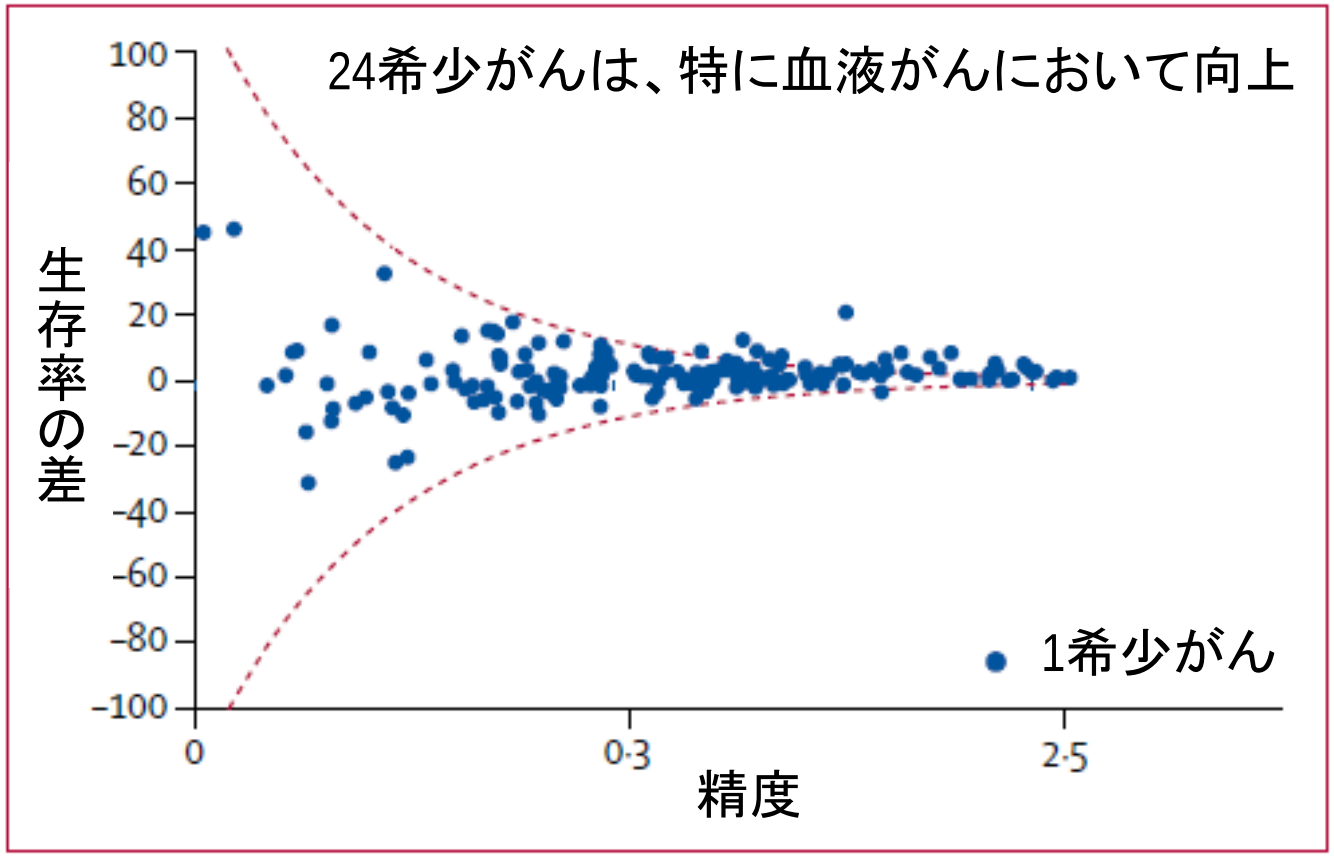
Interpretation Our study benefits from the largest pool of population-based registries to estimate incidence and survival of about 200 rare cancers. Incidence trends can be explained by changes in known risk factors, improved diagnosis, and registration problems. Survival could be improved by early diagnosis, new treatments, and improved case management. The centralisation of treatment could be improved in the seven European countries we studied.

希少がんの5年相対生存率（RS%）の推計（ヨーロッパ、診断年2000-2007；追跡2008）

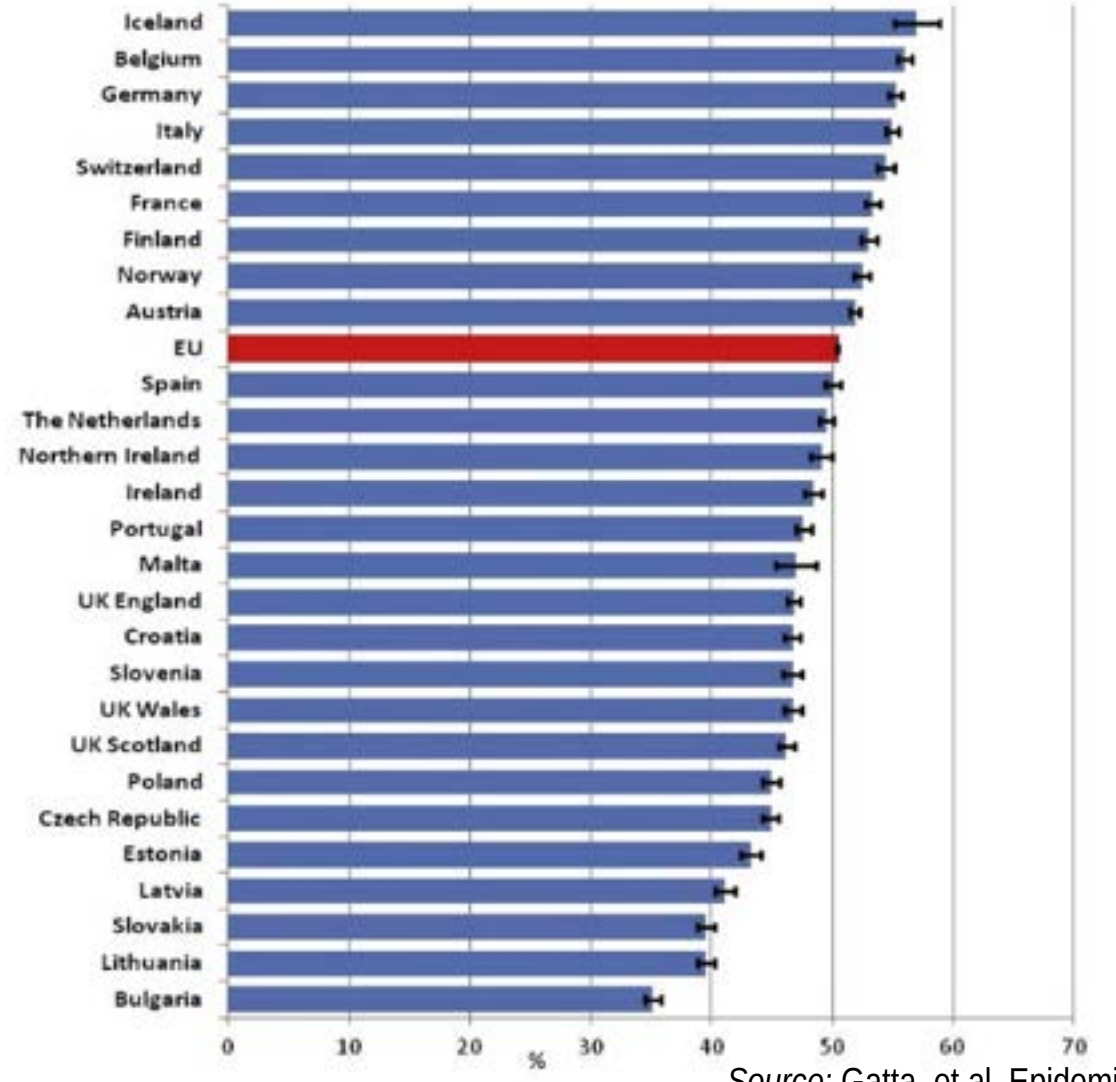
	5年相対生存率
希少胸部がん	13·4%
希少消化器系がん	15·3%
中枢神経がん	21·3%
希少血液がん	50·5%
希少頭頸部がん	52·1%
希少神経内分泌腫瘍	53·5%
希少女性生殖器がん	57·7%
骨肉腫	58·6%
軟部肉腫	59·5%
希少皮膚がん、皮膚以外のメラノーマ	70·2%
起床男性生殖器がん、泌尿器がん	73·6%
希少胎児性腫瘍	78·6%
内分泌器官の希少がん	88·1%

Source: Gatta et al. Burden and centralised treatment in Europe of rare tumours: results of RARECAREnet—a population-based study. Lancet Oncol 2017; 18: 1022–39

希少がんの5年相対生存率の差（1999–2001対2005–07）



年齢とリスクで補正された5年相対生存率（%）全希少がん　ヨーロッパ国別及びEU（診断 2000-2007；追跡2008）エラーバーは95%信頼区間



Source: Gatta et al. Epidemiology of rare cancers and inequalities in oncologic outcomes. EJSO, 2017 in press

生存率の差？

- 診断時ステージ
  - 医療へのアクセス
  - 社会経済状況
  - 人種
  - がん検診
  - 画像診断を含む診断へのアクセス
- 治療
  - 医療へのアクセス
  - 社会経済状況
  - 人種
  - 革新的薬剤
  - 放射線治療施設へのアクセス

Burden and centralised treatment in Europe of rare tumours: results of RARECAREnet—a population-based study

*Population in millions. Number of hospitals providing 75% of treatments (H75), mean annual number of treatments (treat) provided by H75 hospitals, by country and cancer group.

	Belgium (10·5*)			Bulgaria (7·7*)			Finland (5·3*)			Ireland (4·2*)			Netherlands (16·3*)		
	Cases	H75	Treat	Cases	H75	Treat	Cases	H75	Treat	Cases	H75	Treat	Cases	H75	Treat
Head and neck	2098	29	105·6	1180	10	145·1	439	6	82·2	368	7	63·0	2439	12	201·4
Epithelial ovary	760	50	19·5	627	16	52·3	370	10	44·5	261	15	21·0	1118	47	30·2
Oesophagus	689	31	29·3	77	14	5·2	163	8	21·6	289	9	37·1	1422	31	42·0
CNS	623	20	48·4	412	13	41·7	57	4	19·1	229	3	106·3	912	14	84·0
Soft tissue sarcoma	500	35	16·6	372	21	18·4	165	7	25·6	157	17	10·6	802	33	26·4
Thyroid	576	34	14·2	220	12	20·4	286	12	22·8	98	11	9·6	418	31	17·1
Testis	244	40	8·4	180	19	12·4	101	9	14·3	144	12	15·6	609	42	18·4
Biliary tract	214	44	4·9	183	23	6·5	147	13	11·3	122	14	7·7	582	38	12·2
Gastroenteric-pancreatic neuroendocrine tumour	287	46	5·6	30	21	1·3	148	13	9·3	61	20	2·7	355	44	6·9
Liver	250	22	11·0	107	12	7·6	165	11	12·8	68	12	4·6	236	36	5·2
Urinary tract	292	48	6·7	67	17	4·1	48	12	3·9	24	10	2·3	419	46	7·7
Mesothelioma	184	25	8·7	34	10	3·7	64	9	6·8	25	11	2·0	481	43	9·8
Vagina	172	35	5·8	120	9	14·0	70	5	14·8	40	9	4·7	296	14	21·8
Bone sarcoma	81	10	10·2	55	13	4·6	28	3	9·6	30	7	5·2	195	5	43·3
Anal canal	95	27	5·3	39	12	4·1	24	7	4·6	30	9	4·4	135	22	7·2
Melanoma of uvea	43	2	21·9	17	7	2·7	6	1	5·5	29	4	5·7	156	2	80·2
Penis	63	43	1·4	39	17	2·4	21	10	2·1	20	15	1·2	109	26	3·7
Small intestine	62	37	1·9	15	13	1·1	26	13	2·1	27	20	1·3	120	38	2·6
Neuroendocrine carcinoma of skin	46	32	1·9	1	3	0·4	0			15	18	0·8	77	37	2·3
Non-epithelial ovary	20	19	1·3	43	17	3·2	8	9	1·1	8	15	0·6	32	24	1·4
Endocrine carcinoma of thyroid	31	22	1·4	10	9	1·2	8	8	1·2	5	10	0·5	32	13	2·7
Thymus	22	20	1·4	7	8	1·3	4	5	1·1	5	5	1·3	36	15	2·8
Nephroblastoma	18	4	7·4	6	3	2·8	8	3	4·7	7	1	13·4	30	4	16·9
Melanoma of mucosa	14	24	0·8	2	5	0·8	10	7	1·7	6	11	0·6	34	13	3·0
Adrenal cortex	13	14	1·1	13	10	1·3	6	7	0·9	5	11	0·4	25	15	1·5
Embryonal CNS	21	9	4·2	14	9	2·5	6	3	3·1	9	3	6·3	0		
Neuroblastoma	15	4	5·7	8	5	1·7	1	1	2·1	7	2	5·4	12	4	6·2
Retinoblastoma	10	1	14·0	3	5	0·5	3	2	1·5	3	2	1·8	22	1	30·7
Trachea	10	18	0·9	5	4	1·1	4	5	0·9	2	4	0·4	11	11	1·1

日本では？

Cancer Epidemiology 38 (2014) 490–495

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Cancer Epidemiology

The International Journal of Cancer Epidemiology, Detection, and Prevention

journal homepage: www.cancerepidemiology.net

The burden of rare cancer in Japan: Application of the RARECARE definition

Tomoko Tamaki [a,1,*], Yiqi Dong [a], Yuko Ohno [a], Tomotaka Sobue [b], Hiroshi Nishimoto [c], Akiko Shibata [c]

[a] Department of Mathematical Health Science, Graduate School of Medicine, Osaka University, 1-7 Yamadaoka, Suita 565-0871, Osaka, Japan
[b] Department of Social and Environmental Medicine, Graduate School of Medicine, Osaka University, 2-2 Yamadaoka, Suita 565-0871, Osaka, Japan
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一般的ながんと希少がんの差（EUと日本）

Entitie of tumor	Incidence rate per 100,000 in Japan	Incidence rate per 100,000 in EU27
Common in Japan and Rare in EU27		
Squamous cell carcinoma with variants of oesophagus	10.58	3.40
Hepatocellular carcinoma of Liver and IBT	23.66	3.09
Adenocarcinoma with variants of gallbladder and EBT	7.5	2.62
Carcinomas of thyroid gland	8.16	3.65
Rare in Japan and Common in EU27		
Invasive lobular carcinoma of breast	1.05	7.18
Adenocarcinoma with variants of corpus uteri	4.98	9.53
Malignant skin melanoma	0.93	12.41
Basal cell carcinoma of skin	3.34	32.05
Squamous cell carcinoma with variants of skin	2.87	16.39

罹患率および希少がんと一般的ながん分布　簡易分類（EUと日本）

Rare or common	Summary sites	Crude incidence per 100,000 per year		Incidence distribution (%)		Estimated incidence cases in Japan per year
		Japan	EU27	Japan	EU27	
Rare	Digestive tract	6.7	17.5	3	15	8,414
Common	Digestive tract	198.5	75.3	77	67	250,012
Other	Digestive tract	NA	NA	20	18	NA
All	Digestive tract	259.3	113.7	100	100	326,556
Rare	Respiratory tract	6.9	13.6	10	21	8,729
Common	Respiratory tract	45.3	31.5	63	49	57,109
Other	Respiratory tract	NA	NA	27	30	NA
All	Respiratory tract	72.0	63.9	100	100	90,714
Rare	Skin	8.2	1.5	100	2	10,301
Common	Skin	0.0	60.8	0	96	0
Other	Skin	NA	NA	0	2	NA
All	Skin	8.2	63.2	100	100	10,303
Rare	Breast	5.1	4.4	14	7	6,404
Common	Breast	24.0	47.5	65	74	34,427
Other	Breast	NA	NA	13	19	NA
All	Breast	37.0	64.1	100	100	46,647
Rare	Female genital tract	16.5	16.1	86	55	20,792
Common	Female genital tract	0.0	9.5	0	32	0
Other	Female genital tract	NA	NA	14	13	NA
All	Female genital tract	19.3	29.5	100	100	24,303
Rare	Male genital tract	1.2	4.4	3	8	1,496
Common	Male genital tract	30.3	40.6	85	78	38,122
Other	Male genital tract	NA	NA	12	14	NA
All	Male genital tract	35.8	51.9	100	100	45,064
Rare	Urinary system	3.5	2.6	13	8	4,446
Common	Urinary system	17.8	25.8	68	78	22,433
Other	Urinary system	NA	NA	19	14	NA
All	Urinary system	26.2	33.0	100	100	32,988
Rare	Haematopoietic system	11.0	15.9	40	72	13,855
Common	Haematopoietic system	12.0	4.8	43	22	15,132
Other	Haematopoietic system	NA	NA	17	6	NA
All	Haematopoietic system	27.8	22.0	100	100	34,978
Rare	All sites	75.2	108.3	15	22	94,768
Common	All sites	336.1	297.4	65	59	423,298
Other	All sites	NA	NA	20	19	NA
All	All sites	513.9	502.1	100	100	647,346

NA = not available.

希少がん（米国、成人）

North American Association of Central Cancer Registries（NAACCR）
39 人口ベースがん登録

RESEARCH ARTICLES

The Occurrer
in U.S. Adult

ROBERT T. GREENLEE, PHD, MPH[a]
MARC T. GOODMAN, PHD, MPH[b]
CHARLES F. LYNCH, MD, PHD[c]
CHARLES E. PLATZ, MD[d]
LORI A. HAVENER, CTR[e]
HOLLY L. HOWE, PHD[f]

SYNOPSIS

Objective. Rare cancers have been traditionally understudied, reducing the progress of research and hindering decisions for patients, physicians, and policy makers. We evaluated the descriptive epidemiology of rare cancers using a large, representative, population-based dataset from cancer registries in the United States.

Methods. We analyzed more than 9 million adult cancers diagnosed from 1995 to 2004 in 39 states and two metropolitan areas using the Cancer in North America (CINA) dataset, which covers approximately 80% of the U.S. population. We applied an accepted cancer classification scheme and a published definition of rare (i.e., fewer than 15 cases per 100,000 per year). We calculated age-adjusted incidence rates and rare/non-rare incidence rate ratios using SEER*Stat software, with analyses stratified by gender, age, race/ethnicity, and histology.

罹患の25%（米国）

- 希少がん<15/100,000/年
- 解剖学に基づくそれぞれの希少がんの部位には多くの組織型を含む（部位ごとに12–43の組織型グループ）

希少がん（ブラジル）

Rare in Europe and common in São Paulo-Brasil	Europe	São Paulo
	Crude Inc rate	Crude Inc rate
Squamous cell carcinoma with variants of larynx	4.61	6.05
Squamous cell carcinoma with variants of oral cavity	3.51	6.25
Squamous ce		9.47
Carcinoma of		9.65
Common in		
Adenocarcino		3.25
Squamous ce		4.22
Poorly differe		1.95
Invasive lobu		5.37
Adenocarcinoma with variants of corpus uteri	9.93	4.75
Renal cell carcinoma with variants	10.08	4.23
Other non Hodgkin, Mature B cell lymphoma	6.37	1.86

罹患の21%（サンパウロ、ブラジル）

Maria Teresa Bustamante Teixeira
Profª Associada do Departamento de Saúde Coletiva
Programa de Pós-graduação em Saúde Coletiva PPgSC-UFJF
Núcleo de Assessoria Treinamento e Estudos em Saúde NATES-UFJF
Universidade Federal de Juiz de Fora -UFJF

希少がんのデータ精度

TJ
ISSN 0300-8916

Tumori 2017; 103(1): 22-32
DOI: 10.5301/tj.5000559

ORIGINAL RESEARCH ARTICLE

Data quality in rare cancers registration: the report of the RARECARE data quality study

Annalisa Trama[1], Rafael Marcos-Gragera[2], Maria Josè Sánchez Pérez[3,4], Jan Maarten van der Zwan[5], Eva Ardanaz[4,6], Christine Bouchardy[7], Juan Manuel Melchor[3], Carmen Martinez[3], Riccardo Capocaccia[8], Massimo Vicentini[9], Sabine Siesling[5,10], Gemma Gatta[1] and the RARECARE working group contributing to the data quality study

希少がんの診断の正確性を検証

38がん登録
13カ国

結論

<u>診断/罹患の完全性</u>

- 全体的に良好（中皮腫、肝血管肉腫、異型慢性骨髄性白血病のより正確な推計）
- レビューにより、一般的なコーディングミスが依然としてあることが判明
  - 胸膜、脳神経系、肝において、転移性のがんが原発としてコード
  - 口腔がんと中咽頭がんとを混同
  - 境界性のカルチノイドを悪性としてコード
- **形態不明の割合が、レビューしたがんの中に多く存在**

助言

- がん登録において、いくつか選んだ希少がんの定期的な内容の再確認
- 登録の質の向上
  - 神経内分泌腫瘍
  - 血液がん
- 病理診断報告中のテキストや情報に、より焦点をあてる
- 詳細な局在と形態を報告する
- **希少がんとその診断の困難さ及びがん登録へ、臨床医、病理医、がん登録担当者の注目を集める**

ESO ADVANCED COURSES, SEMINARS AND SYMPOSIA

31/03/11

RARE SOLID CANCERS: AN INTRODUCTION

31 March - 1 April 2011
Stresa, Italy

Chairs: P.G. Casali, IT - G. Gatta, IT

In collaboration with

RARECARE
Surveillance of Rare Cancers in Europe

STRESA

DESCRIPTION | FACULTY | INFORMATION | PROGRAMME | SLIDES

ESO-ESMO-RCE Clinical Update on Rare Adult Solid Cancers
25/11/2016 - 27/11/2016, Milan, Italy

Chairs: P.G. Casali, IT - R.A. Stahel, CH

Rare cancers make up as many as one fifth of all new cancer cases. Amongst them, pediatric cancers and rare hematologic neoplasms are often covered by dedicated educational events, while adult solid rare cancers are hardly grouped together and approached as a collective group of neoplasms. Indeed, they account for as many as 15% of all new cancer cases, thus being the main group within rare cancers. This Course is aimed at dealing with them on an annual basis, to provide the community of clinical oncologists specialising in these tumors with a regular update of recent advances in this specific area. The ultimate goal is to strengthen the educational coverage of a group of cancers which may be neglected in spite of their collective incidence.

According to the RARECARE project, which provided a definition and a list of rare cancers, rare adult solid cancers include the following big families of tumors, each of them being therefore covered by a correspondingeducational session within this Course:

ESO-ESMO-RCE Clinical Update on Rare Adult Solid Cancers 2017

ESO-ESMO-RCE
CLINICAL UPDATE ON
RARE ADULT SOLID CANCERS
2-4 December 2017
Milan, Italy

Held in collaboration with:

Milan, Italy - 02 Dec - 04 Dec 2017

Rare cancers make up as many as one fifth of all new cancer cases. Paediatric cancers and rare haematological neoplasms are often covered by dedicated educational events while rare adult solid cancers are hardly grouped together as a collective group of neoplasms.

This ESO-ESMO-RCE joint event is aimed at dealing with them on an annual basis, to provide the community of clinical oncologists specialising in these tumours with a regular update of recent advances in this specific area. The ultimate goal is to strengthen the educational coverage of a group of cancers which may be neglected in spite of their collective incidence.

希少がんをどのようにあつかうべきか？

希少がん

臨床的判断（エビデンスの不足）

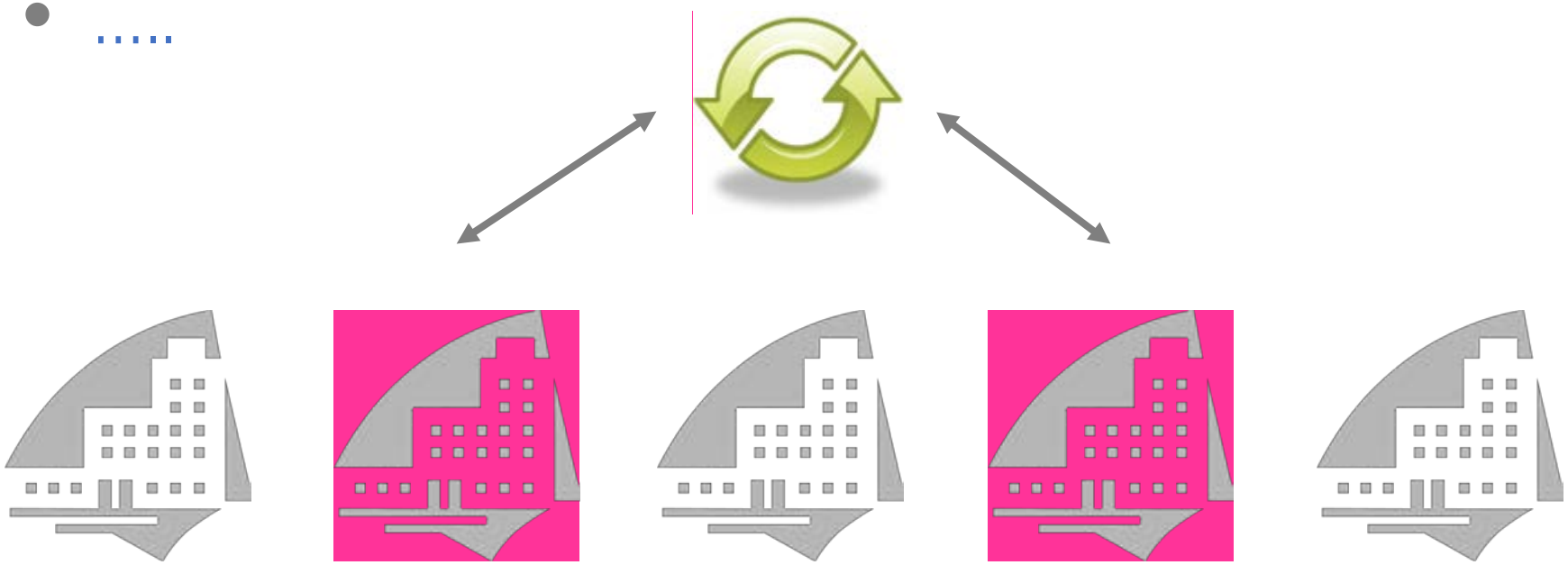
保健医療ケア政策（専門家はどこにいるか？）

臨床研究（少ない患者数）

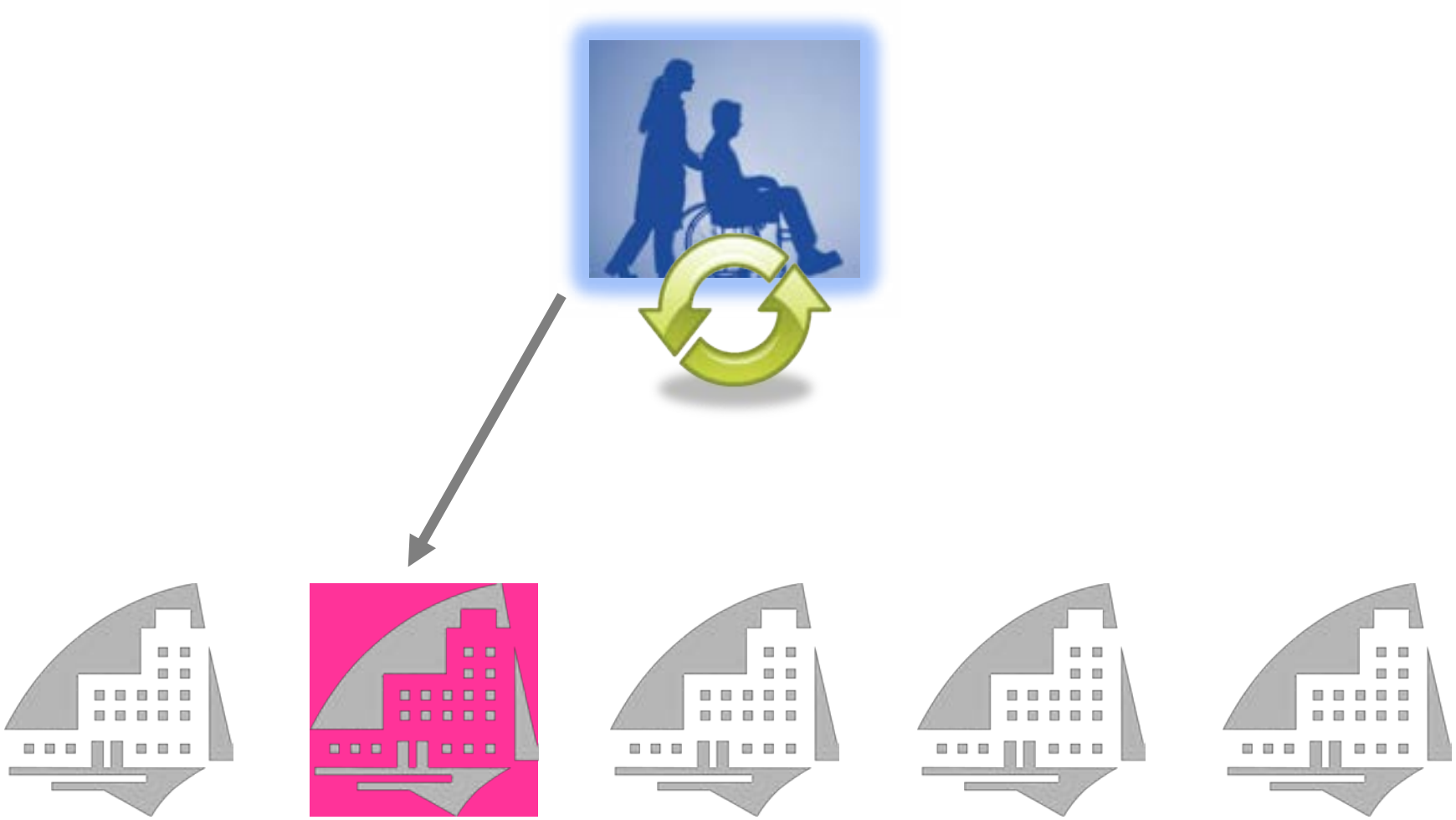
臨床
ネットワーク

遠隔診療

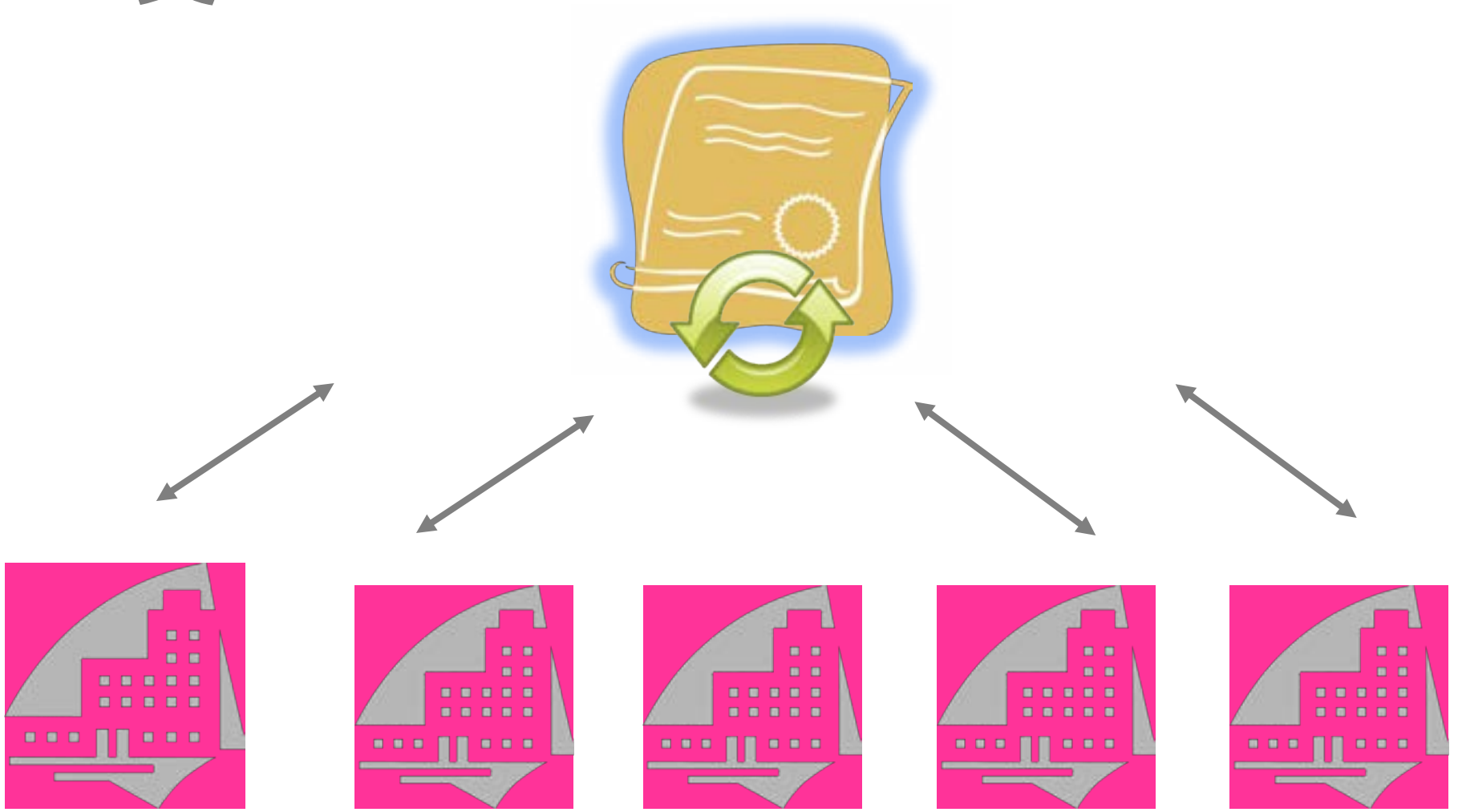
- 病理診断
- 集学的で戦略的な臨床的判断
- 継続した症例情報の共有
- .....



患者の紹介

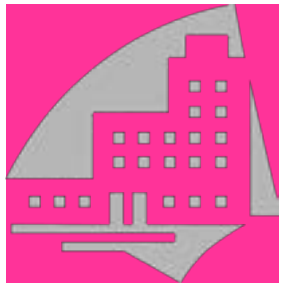


ケアの質



医療者と患者の教育



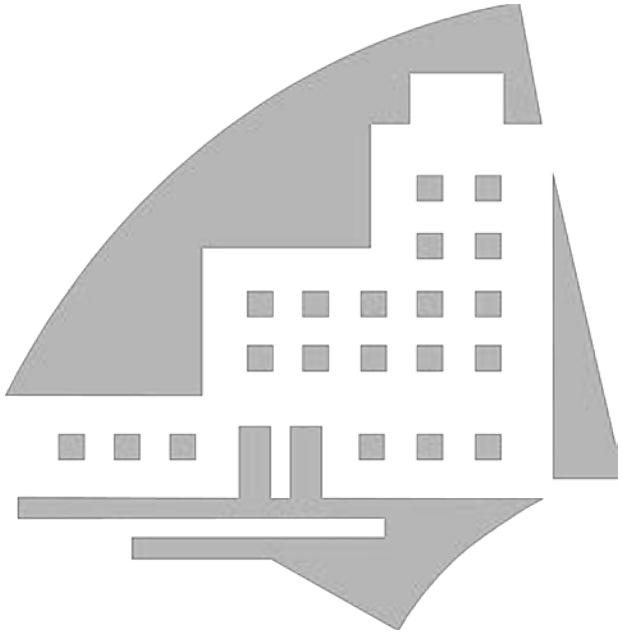


研究のサポート

- 観察研究・がん登録
- バーチャルバイオバンク
- 臨床試験

……

専門家の所属機関から...



…参照ネットワークへ

Show your support and sign the
Call to Action Against Rare Cancers:

www.rarecancerseurope.org

RARE CANCERS

More common than you think!

Rare Cancers Europe is a joint initiative based on a partnership between the European Society for Medical Oncology (ESMO), the European Organisation for Rare Diseases (EURORDIS), the European Cancer Patient Coalition (ECPC), the European Organisation for Research and Treatment of Cancer (EORTC), Conticanet, EuroBoNeT, the World Sarcoma Network (WSN), the Association of European Cancer Leagues (ECL), the Chronic Myeloid Leukaemia Support Group, the International Brain Tumour Alliance (IBTA), Orphanet, the Chronic Myeloid Leukaemia Advocates Network, the Sarcoma Patients EuroNet Association (SPAEN), GIST Support UK & PAWS-GIST, Cancer 52, the International Kidney Cancer Coalition (IKCC), the Chordoma Foundation, the Fondazione IRCCS Istituto Nazionale dei Tumori, the European Institute of Oncology (IEO), the European Society for Paediatric Oncology (SIOP Europe), the European Society of Surgical Oncology (ESSO), the Grupo Español de Tumores Huérfanos e Infrecuentes (GETHI), the European School of Oncology (ESO), the European Oncology Nursing Society (EONS), *e*cancer, the European Society of Pathology (ESP), the European, Middle Eastern and African Society for Biopreservation and Biobanking (ESBB), Novartis Oncology (initiating sponsor and industry partner), Pfizer Oncology (industry partner), and Sanofi (industry partner). The campaign is moreover supported by additional corporate supporters, including Amgen (silver industry supporter) and Takeda Pharmaceuticals Europe (silver industry supporter).


ESMO
GOOD SCIENCE
BETTER MEDICINE
BEST PRACTICE
European Society for Medical Oncology

orphanet

EUROPEAN
CANCER
PATIENT
COALITION

CHORDOMA
FOUNDATION

NOVARTIS
ONCOLOGY

SIOPE
SIOP Europe

Fondazione IRCCS
Istituto Nazionale
dei Tumori

EURORDIS
Rare Diseases Europe

RARER
cancers
FOUNDATION

Pfizer
Oncology

esso

IEO
Istituto Europeo
di Oncologia

ECL

IKCC

Bristol-Myers Squibb

UNIVERSITÀ DEGLI STUDI
DI MILANO

European
Society of
Pathology

IBTA
INTERNATIONAL
BRAIN TUMOUR
ALLIANCE

gist
support uk

Lilly

ecancer
the leading oncology channel

EUROPEAN ONCOLOGY
NURSING SOCIETY



Gethi


European, Middle Eastern & African
Society for
Biopreservation &
Biobanking

cmlsupport

LAM
FEDERATION

EORTC

euroSARC

Sarcoma
Patients
EuroNet

cancer52
the common voice
for less common cancers


RARE CANCERS EUROPE
Joining forces for action
Political Recommendations on Rare Cancers
Download files
ESMO
Improving Rare Cancer Care in Europe
Recommendations on Stakeholder Actions and Public Policies
Whereas

17.

地域レベルで、国レベルで、ヨーロッパレベルでの専門家をヨーロッパ参照ネットワーク（ERNs）に統合を進め、患者の権利の適用について、欧州委員会の提案した指令による特定基準に示されたように、国境を超えた医療では、適切な組織による、効率のよい臨床研究と研究データの臨床への早期移転が必要だ。このようにして、希少がんの臨床管理が推進されるのである。

DIRECTIVES

DIRECTIVE 2011/24/EU OF THE EUROPEAN PARLIAMENT AND OF THE COUNCIL

of 9 March 2011

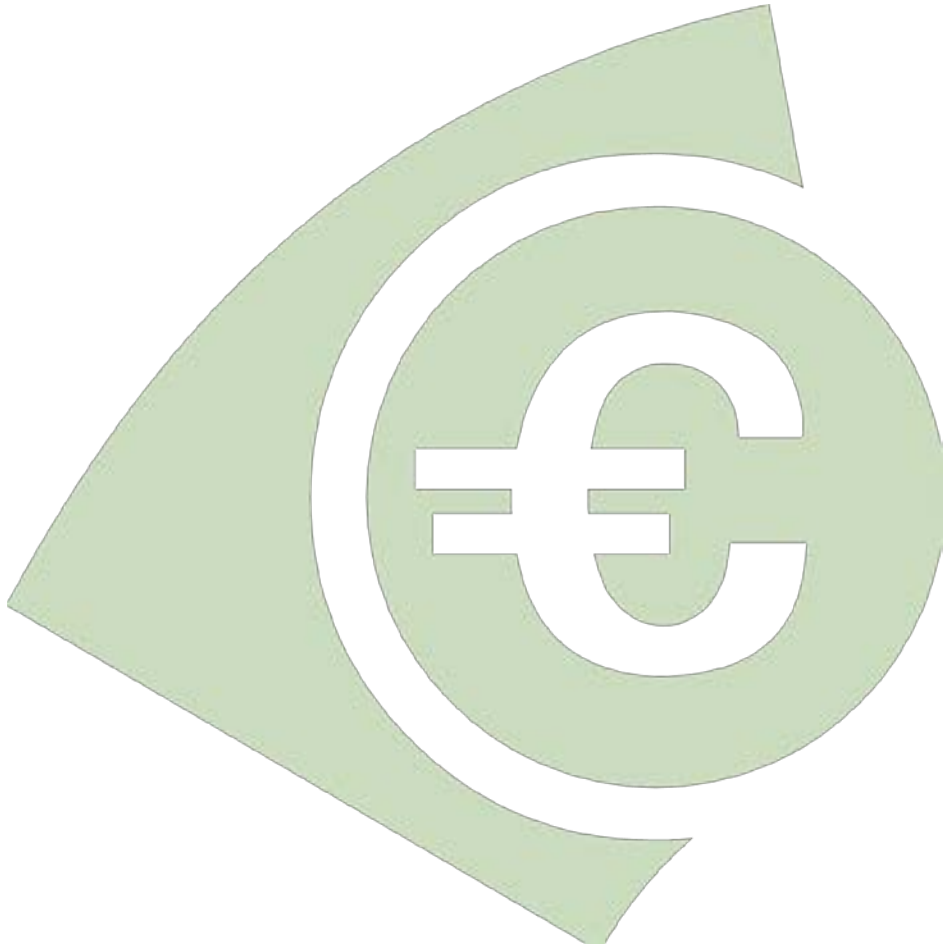
on the application of patients' rights in cross-border healthcare

「希少なまたは有病数の少ない複雑な病気や状態に対し、特別な医療を提供する」

- 的確な診断、治療、フォローアップ、患者の管理をERNsを通じてすることで、高品質で安全な患者へのケアを推進する
- **患者と協働し**、力添え（empower）をする
- 複雑な症例に対して集学的な助言をすることを推奨する
- **臨床ガイドライン**、施設や分野を超えたクリニカルパスを作成し、活用する
- ERNs内外で根拠、専門的意見、**知見を交換し、**集め発信する
- ERNsの中で、共同**研究**を推進する
- 共有できるがん登録を設置することで研究と**疫学サーベイランス**を強化する
- **国立研究所と**ERNsのサポートにより知見とベストプラクティスを交換する

連携

医療業務量




JARC
JOINT ACTION ON RARE CANCERS

JARC
JOINT ACTION ON RARE CANCERS

- 18メンバー国
- 34関連パートナー

協力パートナー

ヨーロッパがん患者連合（ECPC）

ロンドン大学小児健康研究所（UCL-ICH）

ヨーロッパ腫瘍学研修所（ESO）

ヨーロッパ医師会（EMA）

ヨーロッパがん登録協議会（ENCR）

共同研究所（JRC）

ベルギーがん登録（BCR）

イタリア国立保健研究所（Istituto Superiore di Sanità）

ヨーロッパ臨床腫瘍学会（ESMO）

高等衛生研究機関（ISS- Rare Best Practice）

ヨーロッパがん治療研究機構（EORTC）

対がん財団

ヨーロッパ がんリーグ協議会（ECL）

ヨーロッパ外科腫瘍学会（ESSO）

小児がんインターナショナル（CCI-ヨーロッパ）

セントアンナ小児がん研究所（CCRI）

希少がんヨーロッパ（RCE）

EFPIA-EuropaBio

ミラノ大学

Hospital Universitario y Politécnico La Fe. GICT-Cáncer IIS La Fe, （CICT）

提言…

EUでは希少がんに関して、改善のために:

1. 疫学サーベイランス
2. ERNsを通じたケアの質
3. ERNsを通じた臨床ガイドライン
4. ERNsを通じたイノベーション
5. ERNsを通じた医療者と患者の教育
6. 医療政策

European Reference Networks

ERN BOND	European Reference Network on bone disorders
ERN CRANIO	European Reference Network on craniofacial anomalies and ear, nose and throat (ENT) disorders
Endo-ERN	European Reference Network on endocrine conditions
ERN EpiCARE	European Reference Network on epilepsies
ERKNet	European Reference Network on kidney diseases
ERN-RND	European Reference Network on neurological disease
ERNICA	European Reference Network on inherited and congenital anomalies
ERN LUNG	European Reference Network on respiratory diseases
ERN Skin	European Reference Network on skin disorders
ERN EURACAN	European Reference Network on adult cancers (solid tumours)
ERN EuroBloodNet	European Reference Network on haematological diseases
ERN eUROGEN	European Reference Network on urogenital diseases and conditions
ERN EURO-NMD	European Reference Network on neuromuscular diseases
ERN EYE	European Reference Network on eye diseases
ERN GENTURIS	European Reference Network on genetic tumour risk syndromes
ERN GUARD-HEART	European Reference Network on diseases of the heart
ERN ITHACA	European Reference Network on congenital malformations and rare intellectual disability
MetabERN	European Reference Network on hereditary metabolic disorders
ERN PaedCan	European Reference Network on paediatric cancer (haemato-oncology)
ERN RARE-LIVER	European Reference Network on hepatological diseases
ERN ReCONNET	European Reference Network on connective tissue and musculoskeletal diseases
ERN RITA	European Reference Network on immunodeficiency, autoinflammatory and autoimmune diseases
ERN TRANSPLANT-CHILD	European Reference Network on Transplantation in Children
VASCERN	European Reference Network on Rare Multisystemic Vascular Diseases

European Commission

European Reference Networks

EpiCARE . BOND
. CRANIO . ENDO .
ERKNet . EYE . ERNICA .
VASCERN . LUNG . RND . SKIN
. EURACAN . GUARD-HEART
. EuroBloodNet . eUROGEN .
GENTURIS . ITHACA . MetabERN
. PaedCan . RARE-LIVER .
ReCONNET . EURO-NMD .
TRANSPLANT-CHILD .
RITA

Share. Care. Cure.

European Reference Networks

EURACAN

SIOPE INCA EORTC OECI ESTRO EUROPEAN CANCER PATIENT COALITION Sarcoma Patients EuroNet

ESMO esso ECCO European CanCer Organisation EURORDIS RARE DISEASES EUROPE IBTA INTERNATIONAL BRAIN TUMOUR ALLIANCE

Melanoma Patient Network ヨーロッパ

成人の希少固形がん

ミッション

EURACAN は、**成人の希少がん**（RARE ADULTCANCERS、RACs）を対象として、世界をリードし、患者中心で持続可能な集学的な研究機関の立ち上げを目標とする

EURACANは66の保健医療提供者、ヨーロッパ17カ国、22の関連パートナー（PAGs、**希少疾患関係者**）を束ねる

目的

- **病理診断**へのアクセスとEU MS全体での関連治療の向上
- 医学**研修プログラム**の立ち上げと**ケアの質**の向上と均てん化
- **患者会グループとの協働**と、教育資料の広範な提供によるサポート
- **患者の紹介**「ロードマップ」の導入と、紹介状のない患者の専門病医院への紹介
- **臨床上のガイドライン**を作成し、定期的に更新するとともに、新しい**トランスレーショナル研究計画**（及び関連ツール　例：国際データベース、腫瘍バンク）を立ち上げ、推進する
- がんのケアや研究に従事している主要な国内・**国際担当者・ネットワーク**その他の希少疾患の関係者と交流する

結論

- 希少がんの定義は、EUにも、日本を含む他国にも当てはまる
- 希少がんはそれほど珍しい疾患ではない
- 多くの希少がんは予後が不良である（腫瘍ステージ、医療政策、薬へのアクセス？）

- 希少がんデータ（詳細な形態情報つき）は常にアップデートされて利用可能なわけではない
- 人口ベースのがん登録は、希少がん研究には素晴らしいソースであるが、国によっては、臨床的な側面は限定されていることがある（腫瘍ステージ、治療、治療施設）
  - 日本の人口ベースのがん登録データにはこうした情報が含まれている。
- がん登録を国際的に標準化すことに努め、「完全な」国際共同研究が罹患率や生存率を正しく比較することができ、他国の対策から学ぶこともできる

ヨーロッパでは

- 希少がんに関するヨーロッパレベル、国レベルのネットワークが確立されている
- 人口ベースデータと臨床データを統合する仕組みが、最小限の希少がん研究のため必要となる

ご静聴ありがとうございました。

RARE CANCERS EUROPE

JARC
JOINT ACTION ON RARE CANCERS