Cancer Epidemiology 38 (2014) 490–495

Contents lists available at ScienceDirect

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
[c] Division of Surveillance, Center for Cancer Control and Information Services, National Cancer Center, 5-1-1 Tsukiji Chuo-ku, Tokyo 104-0045, Japan

ARTICLE INFO

Article history:
Received 25 June 2014
Accepted 30 July 2014
Available online 22 August 2014

Keywords:
Rare cancers
Incidence
Cancer registries
Population-based

ABSTRACT

Background: Despite the fact that rare cancer is a new target of cancer control in Japan, the incidence of rare cancers is unknown and there is no generally accepted definition of rare cancers in this country. With the aim of calculating incidences of rare cancers in Japan, we therefore adopted a definition and classification of rare cancers that had been published in the European Union (EU) in 2011.
Methods: Using incidence data between 1998 and 2007 submitted by 12 of population based cancer registries in Japan that met our quality criteria and drawing on the EU definition (incidence <6 per 100,000 per year), we estimated the incidences of 845 combinations of tumor sites and histological groups and thus identified the cancers that are rare in Japan.
Results: After identifying 193 combinations of tumor sites and histological groups that fit our criteria for rare cancers, we estimated their incidence to be about 75 per 100,000, which corresponds to about 94,800 new diagnoses in 2012 or approximately 15% of all cancer diagnoses. The categorization of rare and common cancers was almost the same in Japan as in EU.
Conclusions: The present study provides an indication of the size of the rare cancer burden in Japan and epidemiological information to explore this. We are expecting further discussion based on our results with stakeholders in order to construct a Japanese definition of rare cancers.

1. Introduction

Rare cancers were mentioned for the first time in official documents in Japan in the 2012 revision of "The Basic Plan to Promote Cancer Control", which is based on the Cancer Control Act. Because some patient advocacy groups believe that patients with rare cancers receive less medical resources and information than patients with more common cancers, they have requested that more attention be paid to rare cancers with a view to improving management of these patients. Although the medical term "rare cancers" is commonly used, we could find no definition of rare cancers in Japan. The authors believe that there are three basic considerations that underpin the definition of rare cancers. The first is the choice of the best indicator for evaluating rarity. The second is the categorization of cancers defined by a combination of numerous cancer sites and histological types. The last is deciding on the threshold for rarity.

We found two generally accepted definitions of rare cancers in publications from other countries. One is the consensus produced in the European Union (EU) in 2011 by the research project named RARECARE [1], and the other the consensus of a workshop in 2007 sponsored by the National Cancer Institute in the United States (US) [2].

In Europe, rare diseases were defined in the 1990s as those with a prevalence of <50 per 100,000; the same criterion is also often used for rare cancers [3]. However, prevalence is not an appropriate measure of disease frequency for conditions like cancer that have a sub-acute clinical course. Therefore, the RARECARE study group, an international consensus group funded

* Corresponding author. Tel.: +81 6 6879 2526; fax: +81 6 6879 2524.
E-mail addresses: tamaki.tomoko.73e@st.kyoto-u.ac.jp (T. Tamaki), luckyer.tou17@sahs.med.osaka-u.ac.jp (Y. Dong), ohno@sahs.med.osaka-u.ac.jp (Y. Ohno), tsobue@envi.med.osaka-u.ac.jp (T. Sobue), hnishimo@ncc.go.jp (H. Nishimoto), akshibat@ncc.go.jp (A. Shibata).
[1] Present address: Department of Human Health Science, Kyoto University Graduate School of Medicine, 53 Shogoin Kawara-Cho Sakyo Ward, Kyoto 6068507, Japan.

http://dx.doi.org/10.1016/j.canep.2014.07.014

by the Executive Agency for Health and Consumers (EAHC) of the European Commission, agreed on incidence as the most appropriate indicator for measuring the frequency of rare cancers [1].

The incidence of a cancer clearly depends on how that entity is categorized. Standard cancer surveillance summaries typically classify more common cancers by their major anatomic sites or by histologically-defined diagnostic groups. Greenlee et al. [2] reported the incidence of rare and non-rare cancers in US by site and histologic grouping. After extensive discussion with stakeholders comprising oncologists, pathologists, cancer epidemiologists, and patients advocacy groups, the RARECARE study group produced a new type of list of cancers; this list is in three hierarchical layers and takes into account "families of tumors" and "tumors that are clinically meaningful" [1].

Furthermore, an optimal threshold of rarity would depend on both a classification of cancer and the social and relative burden of the diseases of interest at that time. According to the EU definition, rare cancers have an annual incidence of less than 6 per 100,000 [1], whereas according to the US definition the required incidence is less than 15 per 100,000 [2].

After reviewing the studies cited above, we concluded that the strategy of the RARECARE study group for achieving consensus on the definition of rare cancers was sound and therefore decided to adapt the RARECARE definition to assess the burden of rare cancer in Japan.

In the hope that this data will be useful for study and research of rare cancers, we have determined the incidence of rare cancers (annual incidence of less than 6 per 100,000) in Japan and compared our findings with those of the RARECARE study [1].

2. Material and methods

2.1. Data sources

We obtained the Monitoring of Cancer Incidence in Japan (MCIJ) 1993–2007 research data file, based on the September 2011 data submitted by 33 population-based cancer registries (CRs) in Japan [4]. For the present study, we limited cases to invasive cancer diagnosed in the latest 10 years (i.e. 1998–2007), and applied exclusion criteria for comparability and quality of data in Cancer Incidence in Five Continents, Volume IX as following; data with more than 20% of death-certificate-only (DCO) cases, ill-defined site cases, age-unknown cases and an overall the percentage of microscopically verified cases (MV%) less than 75%. [5]. However, we could not evaluate the percentage of age-unknown cases, because the MCIJ data file did not include such cases. The term "cancer" refers to any invasive malignancy, regardless of site or histologic type.

2.2. List of cancers

The present analysis was based on a list of cancer entities produced by the RARECARE study group. We obtained a spreadsheet named "Rare_Cancers_list_March2011" which included information on all cancers defined by the RARECARE project through the website [6]. This list is hierarchically organized into three layers based on various combinations of the third edition of the International Classification of Diseases for Oncology (ICD-O-3) morphology and topography codes [7]. The first layer comprises 59 cancer entities. All cancer entities categorized as first layer are considered to involve the same clinical expertise and patient referral structure. Each first layer entity is split into varying numbers of second layers, of which there are 197 entities; all second layer entities are considered to require similar clinical management and research. Each second layer entity is further divided into varying numbers of third layers; these correspond to the World Health Organization names of individual cancer entities and their corresponding ICD-O-3 codes. There are 587 cancer entities in the third layer.

In our Japanese version, we added two cancer entities (morphology code 8700; pheochromocytoma, malignant and morphology code 8710-8711; Glomus tumor, malignant) to the second layer under the first layer of neuroendocrine tumors and non-glial tumors of cranial and peripheral nerves, autonomic nervous system and paraganglia, respectively. Therefore, the second layer comprises 197 entities in the original list, but 199 entities in our study.

2.3. Incidence

Based on the list of cancers mentioned above, we calculated incidences of each cancer entities (total of 845) and all cancer combined, as the number of new cases occurring in 1998–2007 divided by the total person-years in the general (male and female) population in each CR region. The total number of new cases occurring in 1998–2007 was approximately 632,400 from 12 selected CRs. The cumulative population covered was about 121 million, which corresponds to 10% of the population in Japan over the period 1998–2007. We estimated the expected number of new cases in Japan in 2012 by assuming that the incidences in all of Japan were the same as those in our sample and multiplying the crude incidence by the 2012 Japanese population (125,957,000). We contrasted the incidences in Japan with those in EU which reported by Gatta et al. [1] or published online by the RARECARE project ("Rare_Cancers_list_March2011") [6].

After calculating incidences, we categorized each cancer entity of the first or second layer into rare or common based on the RARECARE definition (incidence $<$6/100,000/year is rare) the same as the RARECARE study [1].

2.4. Data quality indicators

We calculated the proportion of DCO cases and MV cases among our subject data. In addition, the following two other data quality indicators were calculated regarding the accuracy of diagnoses and completeness of incidence for rare cancers; the proportion of cases with not-otherwise-specified (NOS) morphology (M) codes (ICD-O-3 M8000 and 8001) and the cases with poorly defined topography (codes C260, C268, C269, C390, C398, C399, C559, C579, C639, C689, C729, C759–765, C767–768).

3. Results

An excerpt of our result regarding incidences for 845 cancer entities in total and the corresponding incidences in EU [6] is shown in Table 1, as a part of online data for information to help understanding structure of the cancer list. Additional data may be found in the online version of this article (Table A).

In Japan there were 16 entities with incidence $>$6/100,000/year (common cancers) and in EU 19 such entities. Conversely, in Japan there were 193 entities with incidence under 6/100,000/year (rare cancers) and in EU 186 such entities. Of the 16 common cancer entities in Japan, four entities were not common in EU, and five cancer entities were common in EU but rare in Japan (Table 2).

About 514 persons per 100,000 were diagnosed with cancer in Japan each year between 1998 and 2007: this translates to 647,300 new cancer cases in 2012. The annual incidence of all rare cancers in Japan was about 75 per 100,000 between 1998 and 2007: this translates to about 94,800 new diagnoses in 2012.

Table 3 shows the incidences of rare and common cancers by summary sites in comparison between Japan and EU [1]. Although the statistics in the article by Gatta et al. [1] and those in the list

Table 1
Incidence rates in Japan and EU27 by the cancer list of the RARECARE[a] study (Excerpt). Source: EU27 (1998–2002) [6].

Rare (R) or common (C)[b]	Layer	Entitie of tumor	Crude rate per 100,000	
			Japan	EU27
	1	Epithelial tumors of nasal cavity and sinuses	0.86	0.44
R	2	Squamous cell carcinoma with variants of nasal cavity and sinuses	0.63	0.31
	3	Squamous carcinoma	0.59	0.24
	3	Verrucous carcinoma	0.00	<0.01
	3	Squamous cell carcinoma spindle cell	0.01	<0.01
	3	Papillary squamous cell carcinoma	0.00	<0.01
	3	Adenosquamous carcinoma	0.00	<0.01
	3	Squamous cell carcinoma, adenoid	0.00	<0.01
	3	Basaloid squamous cell carcinoma	0.00	<0.01
R	2	Lymphoepithelial carcinoma of nasal cavity and sinuses	0.00	<0.01
R	2	Undifferentiated carcinoma of nasal cavity and sinuses	0.03	0.02
R	2	Intestinal type adenocarcinoma of nasal cavity and sinuses	0.00	<0.01
	1	Epithelial tumors of nasopharynx	0.45	0.44
R	2	Squamous cell carcinoma with variants of nasopharynx	0.36	0.33
	3	Squamous carcinoma	0.23	0.16
	3	Squamous cell carcinoma nonkeratinizing, NOS	0.02	0.02
	3	Squamous cell carcinoma keratinizing, NOS	0.00	0.02
	3	Papillary squamous cell carcinoma	0.00	<0.01
	3	Basaloid squamous cell carcinoma	0.00	NE
	3	Squamous cell carcinoma, adenoid	0.00	<0.01
	3	Lymphoepithelial carcinoma	0.05	0.06
	3	Undifferentiated carcinoma	0.05	0.06
R	2	Papillary adenocarcinoma of nasopharynx	0.00	<0.01
	1	Epithelial tumors of major salivary glands and salivary-gland type tumors	1.04	1.31
R	2	Epithelial tumors of major salivary glands	0.66	0.73
	3	Squamous carcinoma	0.07	0.12
	3	Large cell carcinoma	0.00	0.01
	3	Lymphoepithelial carcinoma	0.00	<0.01
	3	Carcinosarcoma, NOS	0.00	<0.01
	3	Adenocarcinoma NOS	0.12	0.14
	3	Clear cell adenocarcinoma, NOS	0.00	<0.01
	3	Mucinous adenocarcinoma	0.01	<0.01
	3	Ductal carcinoma	0.04	0.01
	3	Oxyphilic adenocarcinoma	0.01	<0.01
	3	Papillary cystadenocarcinoma, NOS	0.00	<0.01
	3	Adenoid cystic carcinoma	0.13	0.12
	3	Mucoepidermoid carcinoma	0.12	0.12
	3	Acinic cell adenocarcinoma	0.06	0.09
	3	Malignant myoepithelioma	0.01	<0.01
	3	Carcinoma in pleomorphic adenoma	0.03	0.02
Omitted below				

[a] RARECARE: The project Surveillance of Rare Cancers in Europa.
[b] Rare or Common: Rare cancer < 6/100,000 and Common cancer ≥ 6/100,000.

Table 2
The differences of common or rare categorization between Japan and EU27. Source: EU27 (1998–2002) [6].

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

published online by the RARECARE project (“Rare_Cancers_list_-March2011”) [6] seemed to be different regarding the hematopoietic system, we referred to the article version the same as the other sites. The proportion of rare and common cancers does not add up to 100% for each cancer site, because some cancers could not be classified as either rare or common cancer entities because information about their morphology was not available. Therefore, the proportion of cancers not classified as any rare or common cancer entity is shown as “other”. The incidence of rare cancers was 15% of that of all cancers in Japan, which is 7% less than that in EU. The proportion of the “other” category varied with site, being highest for respiratory cancers (27%) and lowest for skin cancers (0%). For all sites, this category was 20% for Japan and 19% for EU; the proportions in the two regions were similar by site except for the hematopoietic system (Japan 17%, EU 6%).

Fig. 1 shows age-specific incidences by age group for rare and common cancers. Rare cancers predominated in persons aged up to 34 years whereas from age 35 years plus, the common cancers became increasingly prominent.

Fig. 2 shows the distribution of number of cancer entities (a) and estimated number of new cancer diagnoses in each year according to incidence (b). About 81% of rare cancers had an annual incidence of under 0.5 per 100,000. These “very rare cancers”, which included most types of rare cancers, accounted for only 16,700 (2.6%) of the 647,300 new cancers diagnosed in each year. Another 17 rare cancer entities with an incidence of 0.5–1 per 100,000 accounted for 15,700 cases (2.4%), whereas the 20 cancer entities with incidence 1–6 per 100,000 accounted for 62,400 cases

Table 3
Incidence and distribution for rare and common cancers by summary site in Japan and EU27. Source: EU27 (1998–2002) [1].

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	647,345

NA = not available.

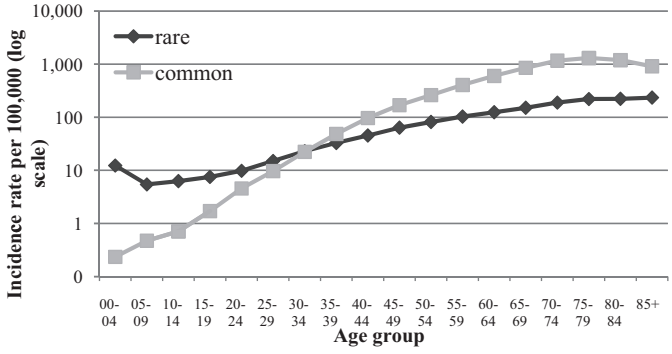

Fig. 1. Age-specific incidence rates per 100,000 for rare and common cancers.

(9.6%) new cases/year in Japan. Sixteen common cancers accounted for the remaining cases.

The data quality indicators regarding our study subject are shown in Table 4. The overall proportion of DCO cases was 9.9%, that of MV cases was 79.1% and that of ill-defined site cases was 1.4%. The overall proportion of cases with "not otherwise specified" morphology codes and that of cases with poorly defined topography was 16.5% and 0.3%, respectively.

4. Discussion

In this study, we comprehensively ascertained the incidence of rare cancers by epidemiological means for the first time in Japan. We calculated incidences for 845 cancer entities in total, and categorized 16 common cancers and 193 rare cancers in Japan based on the published classification of cancers and definition of rarity in EU [1,6]. The categorization of rare and common cancers was almost the same in Japan as in EU. Most of the disparities in cancer incidence pattern between Japan and EU have previously been identified. According to previous studies [8,9], there is a higher incidence of liver cancer, cancer of the gallbladder and extra-hepatic biliary duct, and thyroid cancer and a lower incidence of cancer of the body of the uterus and skin cancer in Japan than in EU: our findings are concordant with this. Previous studies have found that squamous cell carcinoma is the leading morphological type of esophageal cancer in Japan [10], whereas it is adenocarcinoma in European countries [11]. In addition, the incidence of invasive lobular carcinoma of the breast is lower in Asian/Pacific Islander women than in non-Hispanic white women in US [12].

Rare cancers predominantly occur in younger subjects, whereas common cancers become increasingly prominent with increasing age in both Japan and EU. This is not unexpected given that the populations of Japan and EU have a relatively small proportion of children and the risk of most cancers increases with age. Considering that all childhood cancers are rare, it may be better to evaluate the burden of rare cancers among adults only. In their descriptive epidemiological study of rare cancers in US, Greenlee et al. [2] evaluated only persons aged 20 years and older.

On the other hand, we did identify some differences between Japan and EU in patterns of incidence of rare cancers. The

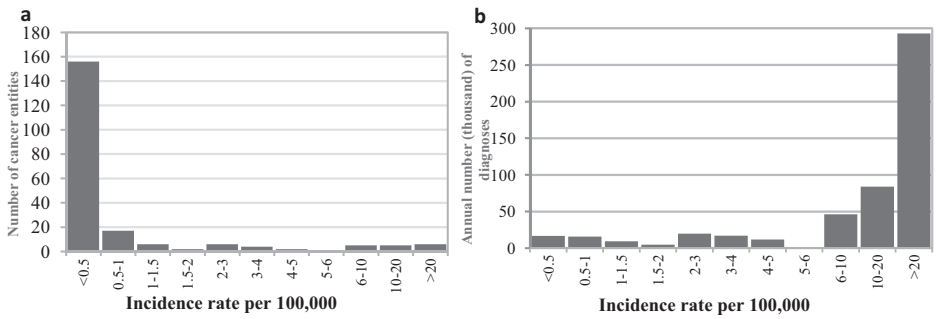

Fig. 2. Distribution of number of cancer entities (a) and annual number of cancer diagnoses (b) in Japan according to categories of incidence rate per 100,000.

Table 4
Data quality indicators of cancers by population-based cancer registry included in the analysis.

CR	Number of cancers	Death certificate only (%)	Microscopic verification (%)	Ill-Defined Cases[a] (%)
CR 1	89,105	9.2	78.7	1.5
CR 2	8,416	5.0	78.1	1.1
CR 3	71,536	9.7	78.8	1.2
CR 4	11,115	15.5	76.8	1.3
CR 5	29,308	9.2	79.6	3.2
CR 6	41,762	3.7	79.3	1.0
CR 7	121,425	16.7	77.4	1.7
CR 8	11,686	12.3	76.0	1.3
CR 9	98,827	7.5	77.7	1.3
CR 10	56,055	6.9	88.9	1.0
CR 11	5,098	6.2	75.8	1.5
CR 12	88,089	8.6	81.8	1.1
All CRs	632,422	9.9	79.1	1.4

[a] Ill-defined: Topology code C26-, C39-, C48-, C76-, C80 CR = cancer registry.

proportion of incidence of rare cancers was 7% lower in Japan as a whole than in EU, in spite of the similar classification of rare and common cancers used in the two studies and the fact that the proportion of incidence of "other" (not categorized as rare or common) was almost the same in Japan as in EU. We do not believe that these findings indicate that there is a smaller burden of rare cancers in Japan than in EU, because the proportion of incidence of all rare cancers or all common cancers varies depending on the incidence of each cancer in any one category. For example, rare cancers accounted for 3% of the incidence of all male genital system cancers in Japan and 8% in EU (Table 3); there is one common cancer and 10 rare cancers in this group of cancers. Thus, in Japan the incidence of each rare cancer in the category of male genital system cancers is lower than that in EU, which may result in lower cumulative incidences of rare cancers in the male genital system in Japan. In contrast, overwhelmingly high incidences of common cancers could lead to those cancers accounting for a greater proportion of the incidence of all common cancers. For example, the leading cancer type in EU is "epithelial tumors of breast", the incidence of which was 64 per 100,000 according to the RARECARE study, whereas in our study the leading cancer type in Japan was "epithelial tumors of stomach", the incidence of which was 94 per 100,000 based on our study, about 5-fold the incidence in EU.

Furthermore, some cancers not classified as either a rare or common cancer entity would affect the incidences of common or rare cancers. For the hematopoietic system, in Japan the proportion of incidence of the "other" category was 17%, which is 3-fold that reported in EU. The difference of main sources of cases for population-based cancer registries would be likely reason for the obvious difference of the proportion of incidence of the "other" category for the hematopoietic system between Japan and EU. Many population-based cancer registries in EU obtain information from hematopoietic laboratories, whereas hospital reports and death certificates are the main sources for most population-based cancer registries in Japan [5]. Therefore, the incidences of both common and rare cancers of the hematopoietic system in our study would be underestimates. Although the proportion of incidence of all rare cancers seems a simple indicator of the size of the burden of rare cancers, it is important to recognize that it can provide misleading information.

Our study had some limitations, one of which being that we did not make an absolute evaluation of the various cancer incidences. Our estimates of incidences are based on the premise that the segments of the population covered by the selected 12 CRs are representative of the population of Japan as a whole. Because we defined most entities of rare cancers according to a combination of specific site and well-defined histology, a combination of poorly defined site and unspecified morphology could not be consistently classified in other way. We therefore decided to include data only from CRs that met our quality criteria, thus excluding CRs with a high proportion of death-certificate only cases and a low proportion of cancers verified by microscopically. This strategy was designed to minimize biasing and underestimating the incidences of rare cancers. However, if we had set our inclusion criteria too strictly, we would not have been able to ensure enough cases to estimate the incidence of rare cancers.

To assess the representativeness of our incidences of common cancers, we compared our data with incidences estimated for all of Japan by the MCIJ 2006 [13]. Our incidences for major cancers (stomach 93.8, colorectal 80.6, lung 68.2, all 513.9) were very similar to those of MCIJ 2006 (stomach 91.5, colorectal 83.7, lung 66.9, all 520.0), suggesting that the population we studied is as representative of the population of Japan as the population covered by the MCIJ 2006.

In conclusion, our estimates indicate that about 94,800 (75 per 100,000 per year) new rare cancers will likely be diagnosed in Japan annually. For the first time, the present study provides an indication of the size of the rare cancer burden in Japan and epidemiological information to explore this. We are expecting further discussion based on our results with stakeholders in order to construct a Japanese definition of rare cancers.

Role of the funding source

The study was supported by the Third-term Comprehensive Ten-year Strategy for Cancer Control from the Ministry of Health, Labour and Welfare (H24-008) (Principal investigator: Tomotaka Sobue).

Conflict of interest statement

The authors have declared no conflicts of interest.

Acknowledgments

The survey on cancer incidence in Japan was conducted with contributions from the 33 prefectural cancer registries: Aomori, Iwate, Miyagi, Akita, Yamagata, Ibaraki, Tochigi, Gunma, Chiba, Kanagawa, Niigata, Toyama, Ishikawa, Fukui, Gifu, Aichi, Shiga, Kyoto, Hyogo, Tottori, Shimane, Okayama, Hiroshima, Yamaguchi, Tokushima, Kagawa, Ehime, Kochi, Saga, Nagasaki, Kumamoto, Kagoshima and Okinawa.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.canep.2014.07.014.

References

[1] Gatta G, van der Zwan JM, Casali PG, Siesling S, Dei Tos AP, Kunkler I, et al. Rare cancers are not so rare: the rare cancer burden in Europe. Eur J Cancer 2011;47:2493–511.
[2] Greenlee RT, Goodman MT, Lynch CF, Platz CE, Havener LA, Howe HL, et al. The occurrence of rare cancers in US adults, 1995–2004. Public Health Rep 2010;125:28–43.
[3] European Parliament and Council of European Communities. Decision no. 1295/1999/EC of the European parliament and of the council of 29 April 1999 adopting a programme of community action on rare disease within the framework for action in the field of public health (1999–2003). European Parliament and Council of European Communities; 1999.
[4] Matsuda T, Matsuda T, Shibata A, Katanoda K, Sobue T, Nishimoto H, et al. Cancer incidence and incidence rates in Japan in 2007: A study of 21 population-based cancer registries for the Monitoring of Cancer Incidence in Japan (MCIJ) Project. Jpn J Clin Oncol 2013;43:328–36.
[5] Curado MP, Edwards B, Shin HR, Storm H, Ferlay J, Heanue M, et al. Cancer incidence in five continents. Vol. IX. Lyon: IARC Scientific Publication, 2008.
[6] The project Surveillance of Rare Cancers in Europa. Rare_Cancers_list_March2011 [Data file]. Retrieved from ⟨http://www.rarecancer.eu/rarecancers/rarecancers.asp⟩ [accessed 15. 1. 14].
[7] Percy C, Fritz A, Jack A, Shanmugaratnam K, Sobin L, Parkin DM, et al. International classification of diseases for the oncology (ICD-O). third ed. Geneva: World Health Organization, 2000.
[8] Ferlay J, Bray F, Pisani P, Parkin DM. GLOBOKAN 2002. Cancer incidence, mortality and prevalence worldwide iarc cancer base No. 5, version 2.0. Lyon: IARC Press, 2004.
[9] Forman D, Bray F, Brewster DH, Gombe Mbalawa C, Kohler B, Piñeros M, et al., eds. Cancer Incidence in Five Continents, Vol. X (electronic version). Lyon: IARC, 2013.
[10] Shibata A, Matsuda T, Ajiki W, Sobue T. Trend in incidence of adenocarcinoma of the esophagus in Japan, 1993–2001. Jpn J Clin Oncol 2008;38:464–8.
[11] Botterweck AA, Schouten LJ, Volovics A, Dorant E, van Den Brandt PA. Trends in incidence of adenocarcinoma of the oesophagus and gastric cardia in ten European countries. Int J Epidemiol 2000;29:645–54.
[12] Hausauer AK, Keegan THM, Chang ET, Clarle CA. Recent breast cancer trends among Asian/Pacific Islander, Hispanic, and African-American women in the US: changes by tumor subtype. Breast Cancer Res 2007;9:R90.
[13] Matsuda T, Marugame T, Kamo K, Kananoda K, Ajiki W, Sobue T, et al. Cancer incidence and incidence rates in Japan in 2006: Based on Data from 15 population-based cancer registries in the monitoring of cancer incidence in Japan (MCIJ) Project. Jpn J Clin Oncol 2012;42:139–47.