希少がんは希少ではない？
ー小児・AYAがんを含めた希少がんの統計

国立がん研究センター
がん対策情報センター
がん統計・総合解析研究部
片野田　耕太

「希少がんはそれほど希少ではない」

- ヨーロッパのRARECAREプロジェクト
- 希少がんを「1年間に人口10万人当たり6例未満しか発生しないがん」と定義
- 希少がんをすべて合わせると1年間に人口10万人当たり108例発生
- がん全体の22%を占める

⇒「希少ではない」

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

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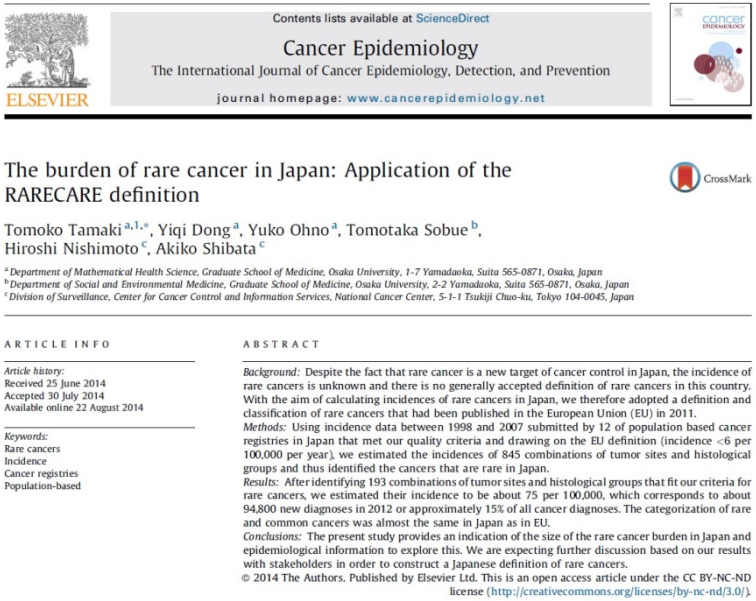
ABSTRACT

Purpose: Epidemiologic information on rare cancers is scarce. The project Surveillance of Rare Cancers in Europe (RARECARE) provides estimates of the incidence, prevalence and survival of rare cancers in Europe based on a new and comprehensive list of these diseases.
Materials and methods: RARECARE analysed population-based cancer registry (CR) data on European patients diagnosed from 1988 to 2002, with vital status information available up to 31st December 2003 (latest date for which most CRs had verified data). The mean population covered was about 162,000,000. Cancer incidence and survival rates for 1995–2002 and prevalence at 1st January 2003 were estimated.
Results: Based on the RARECARE definition (incidence <6/100,000/year), the estimated annual incidence rate of all rare cancers in Europe was about 108 per 100,000, corresponding to 541,000 new diagnoses annually or 22% of all cancer diagnoses. Five-year relative survival was on average worse for rare cancers (47%) than common cancers (65%). About 4,300,000 patients are living today in the European Union with a diagnosis of a rare cancer, 24% of the total cancer prevalence.

日本版RARE CARE

- ヨーロッパのRARECAREと同様の手法
- 精度基準を満たした12の県のがん登録データ
- 希少がんをすべて合わせると1年間に人口10万人当たり75例発生
- がん全体の15%を占める（胃がんと同じ割合）

⇒「希少ではない」



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

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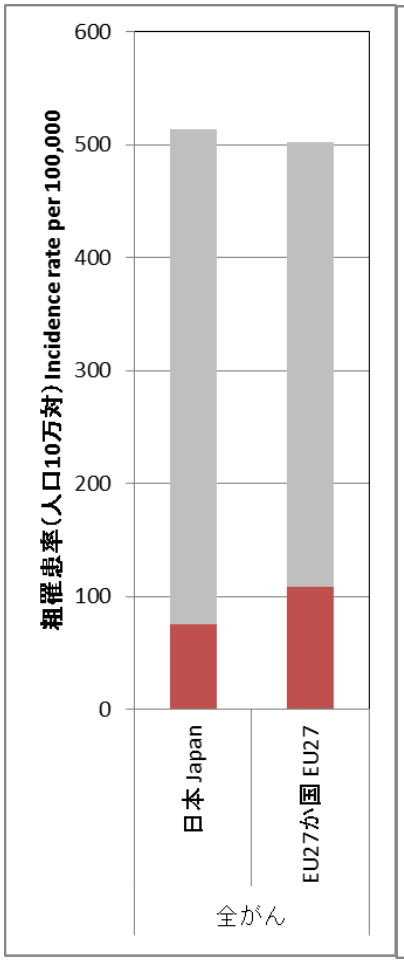
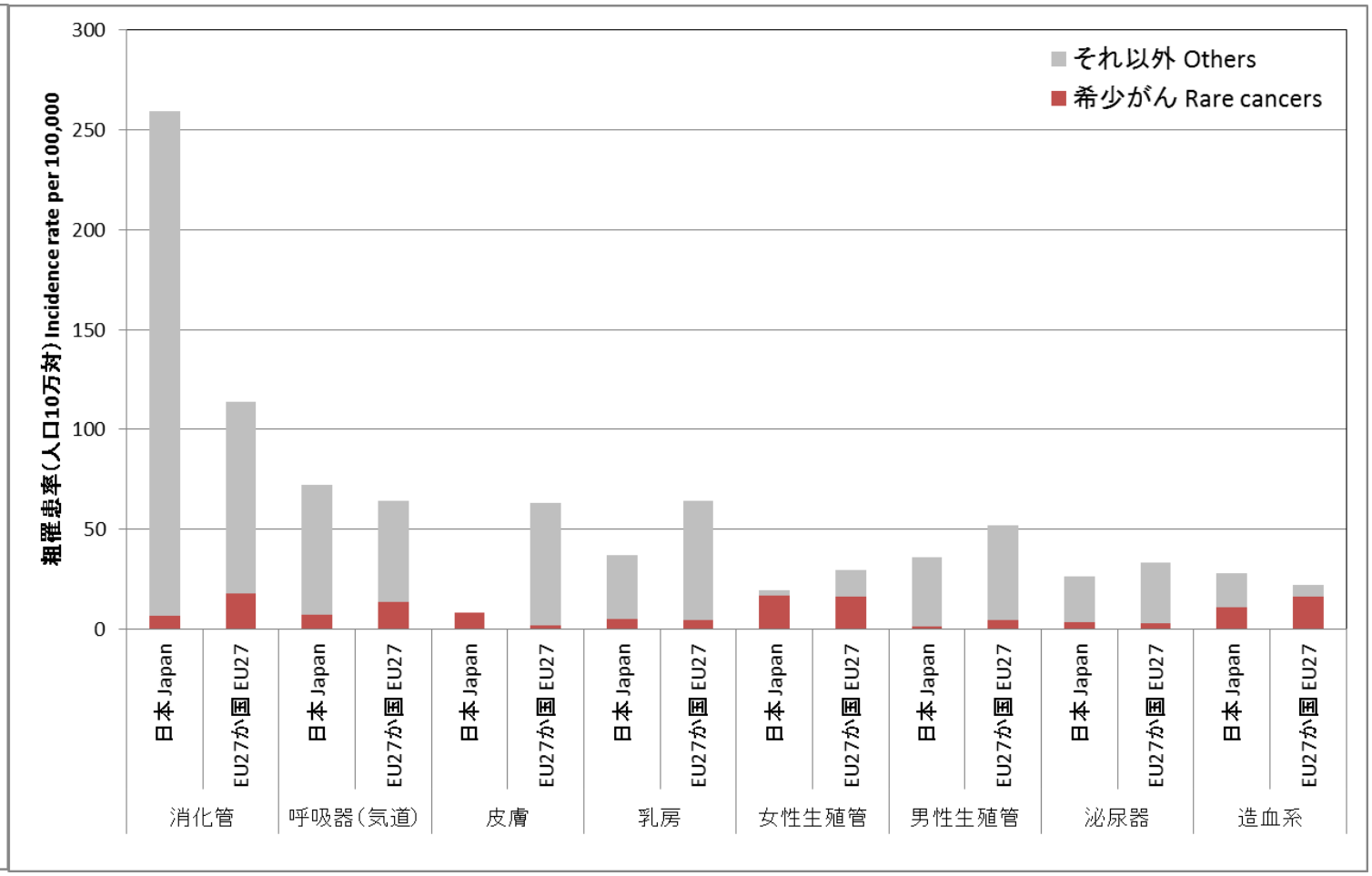
ABSTRACT

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.

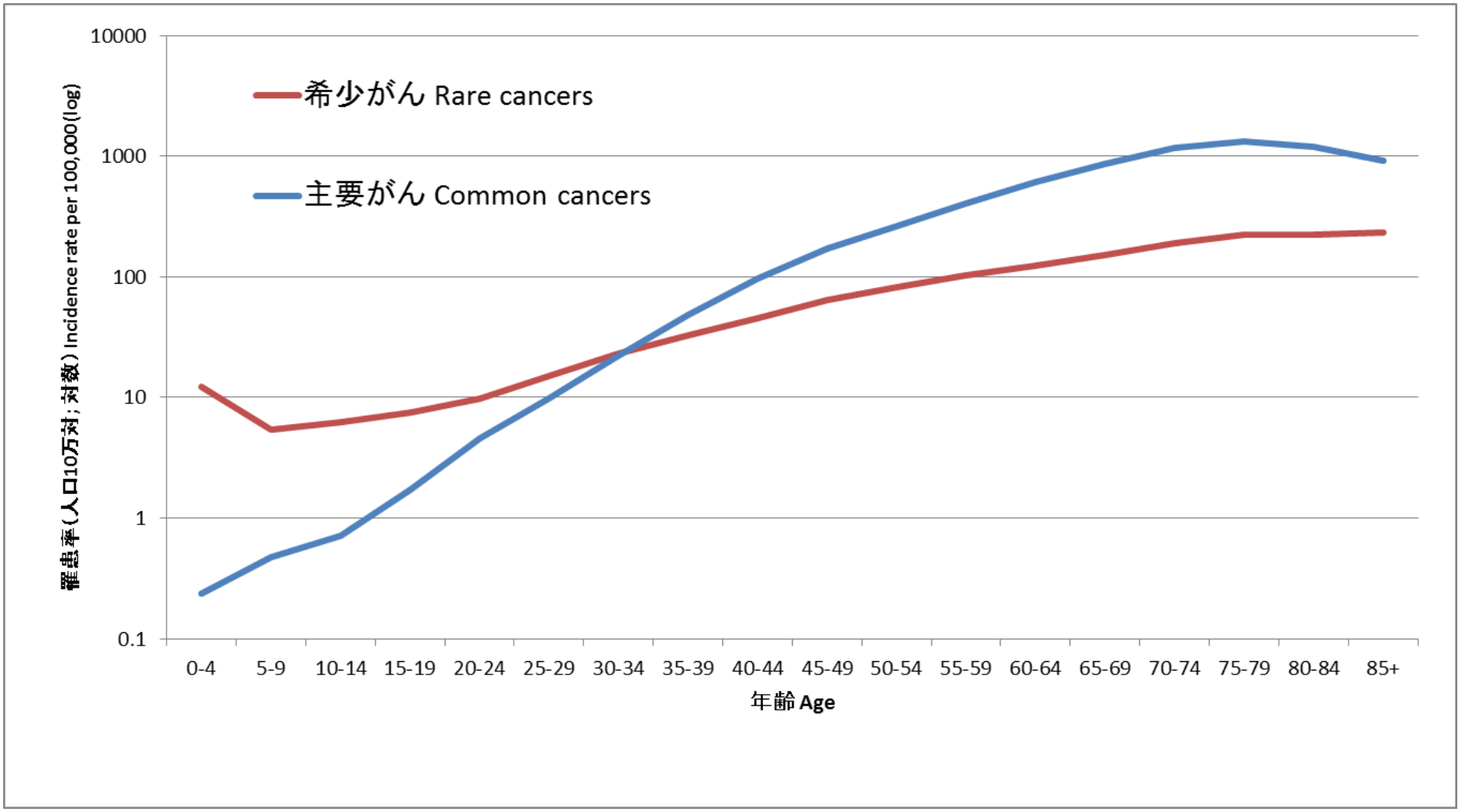
がん種によって「希少がん」の割合は異なる





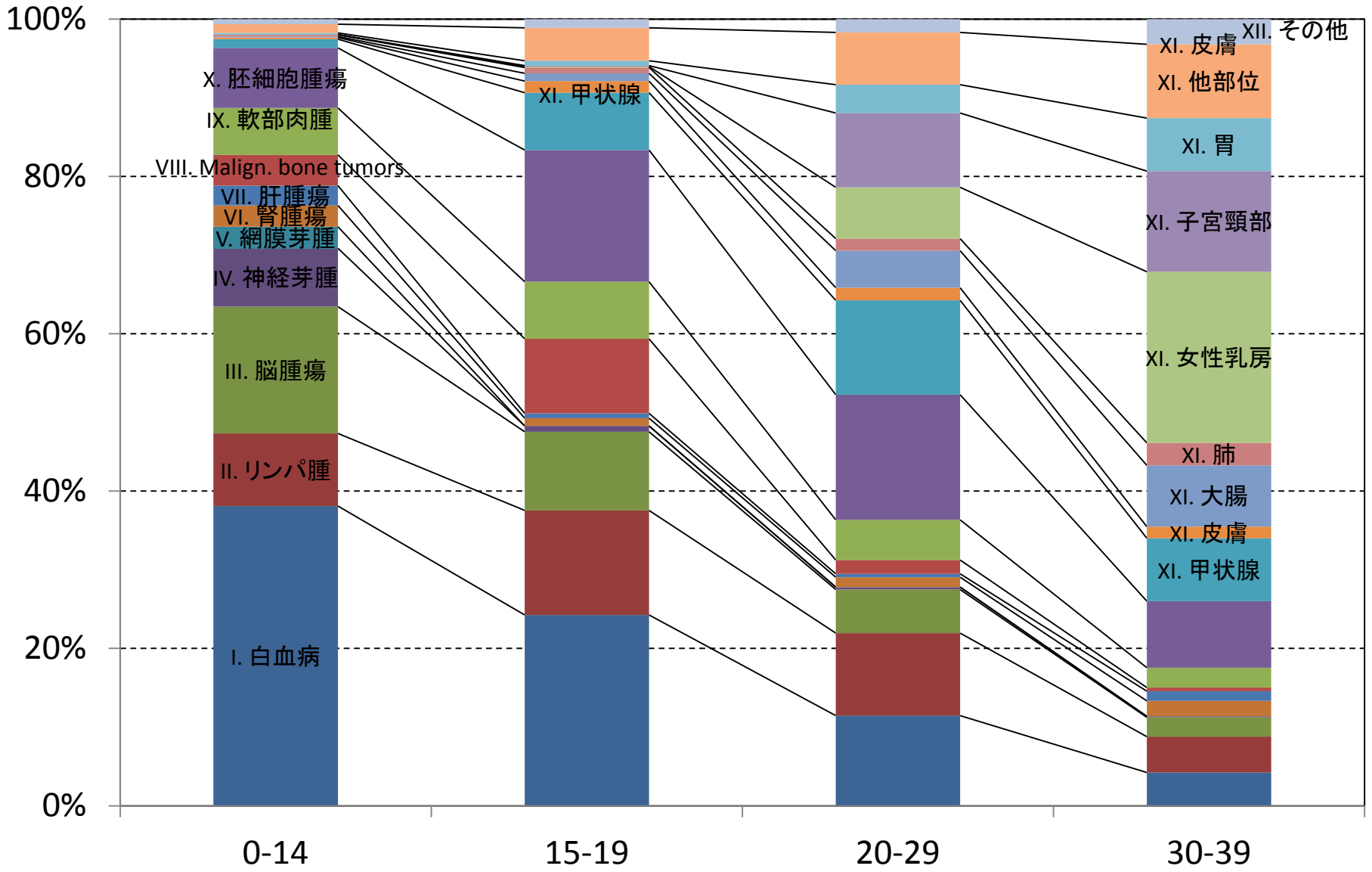
出典： Eur J Cancer 2011; 47: 2493-511
Cancer Epidemiology 2014; 38: 490-5

年齢によって「希少がん」の割合は異なる



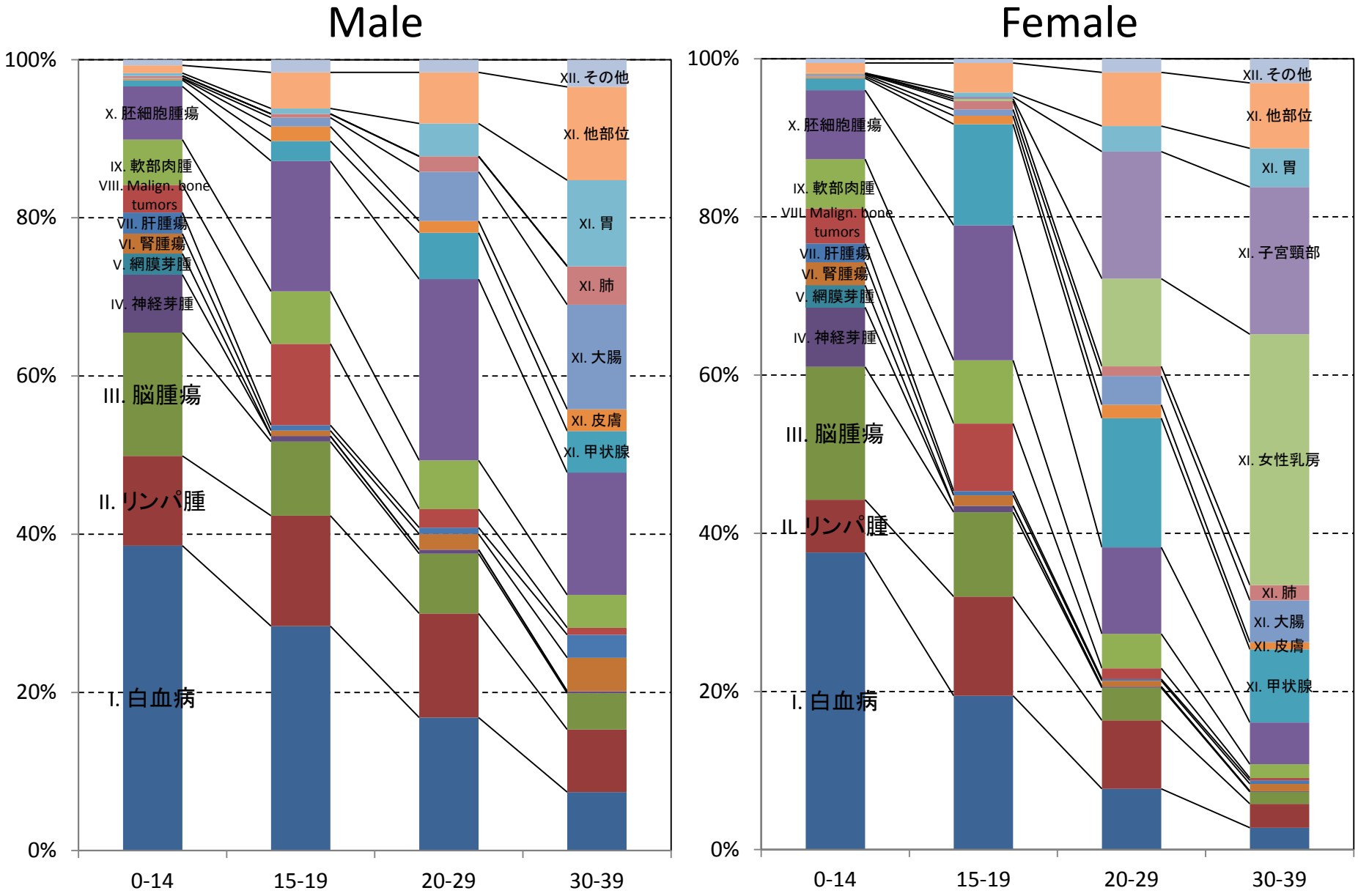
出典： Cancer Epidemiology 2014; 38: 490-5

小児からAYA世代へのがん種の変化



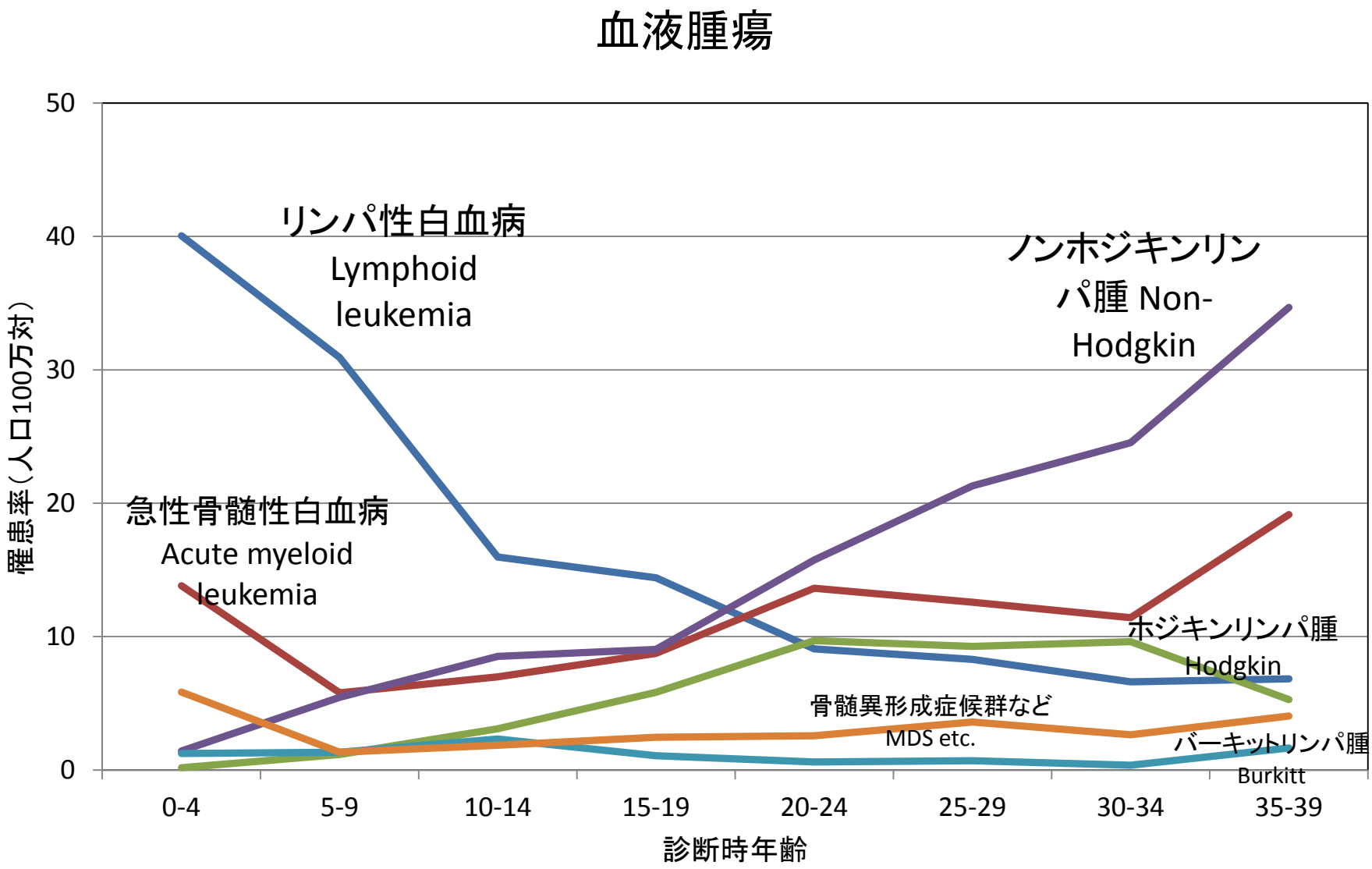
* 悪性腫瘍のみ（脳腫瘍も）　AYA: adolescent and young adult　Katanoda et al. Jpn J Clin Oncol 2017; 47: 762-71（以下同じ）

特に女性でがん種の変化が大きい



* 悪性腫瘍のみ（脳腫瘍も）

がん種別に見た罹患率の年齢変化



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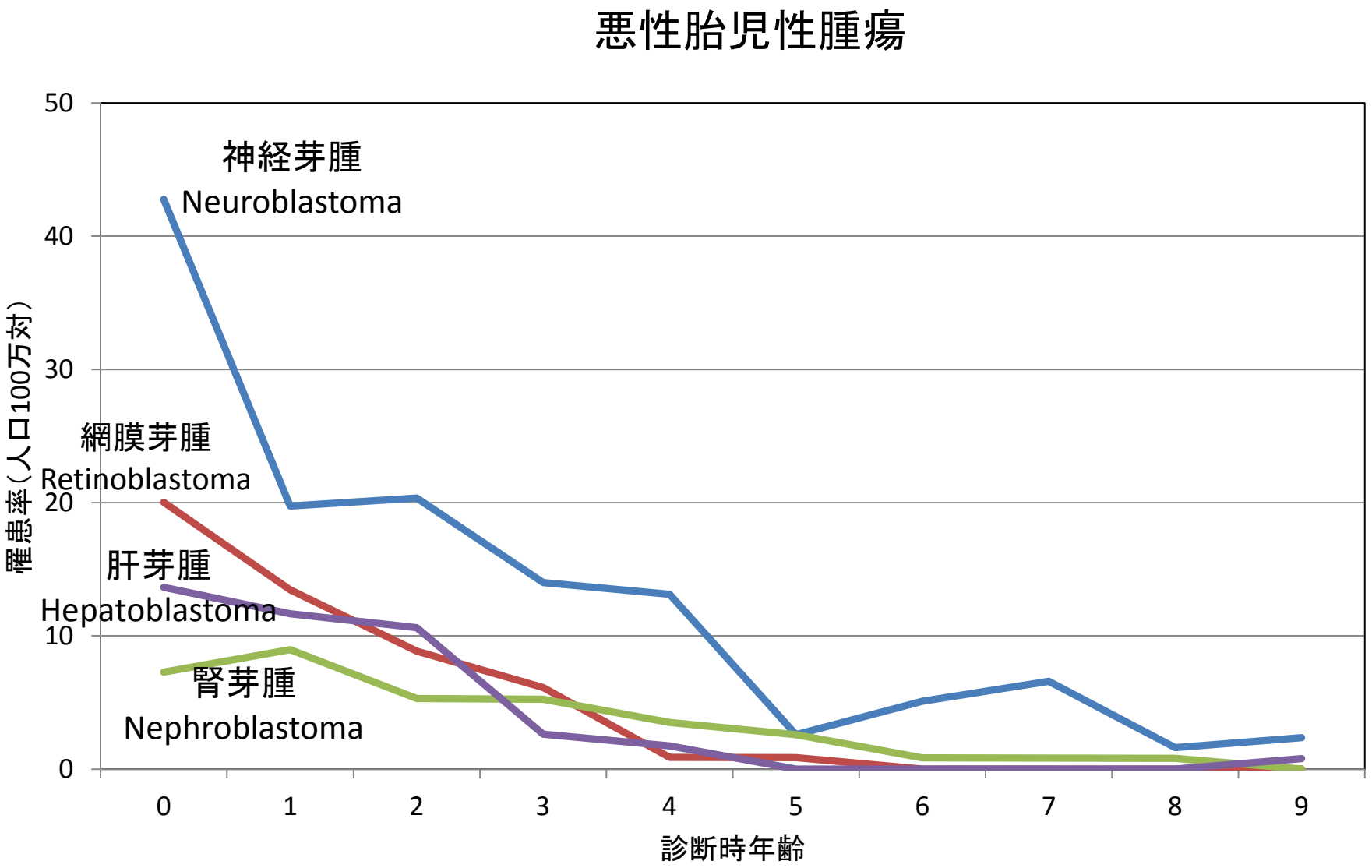

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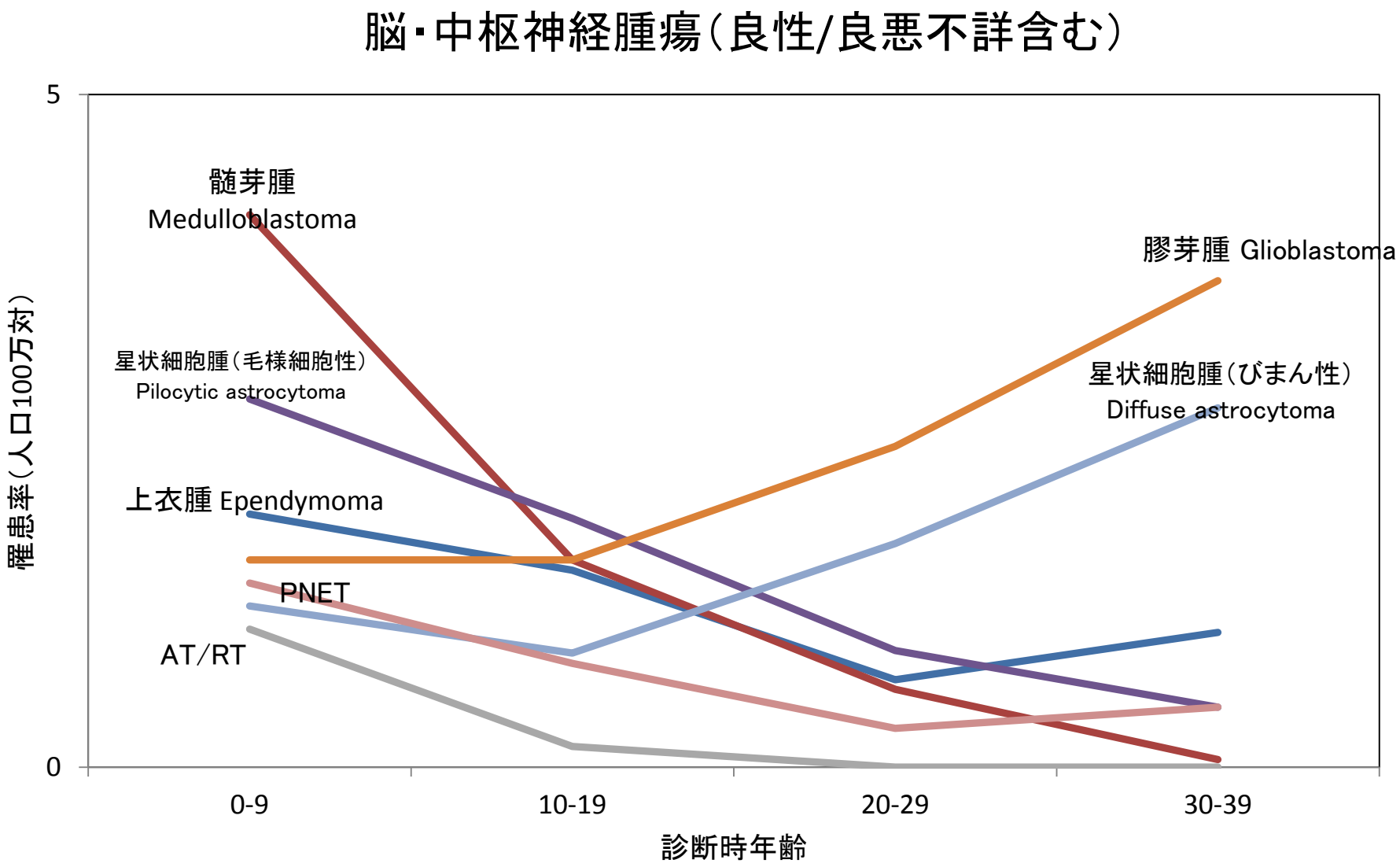
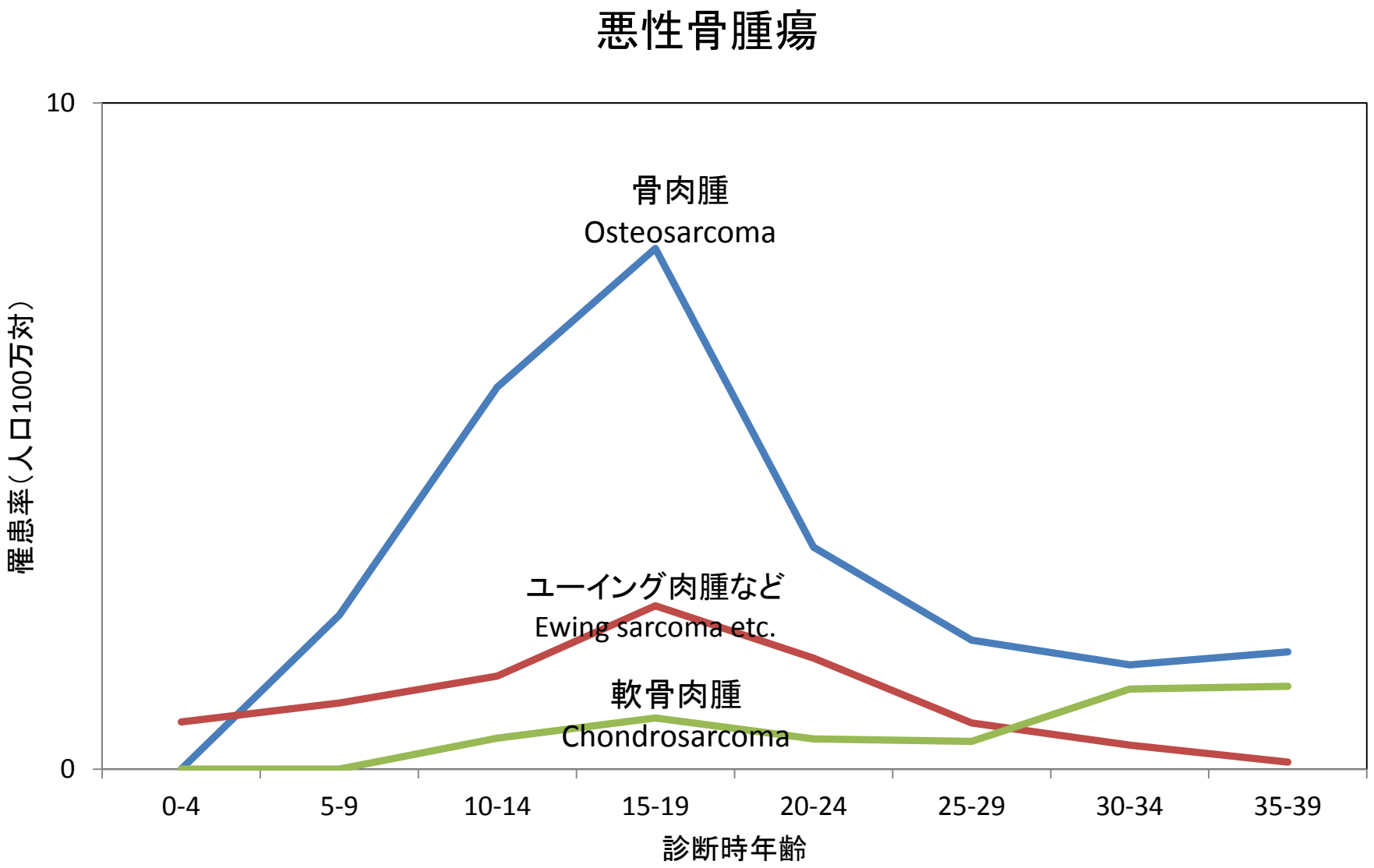


Katanoda et al. Jpn J Clin Oncol 2017; 47: 762-71

がん種別に見た罹患率の年齢変化



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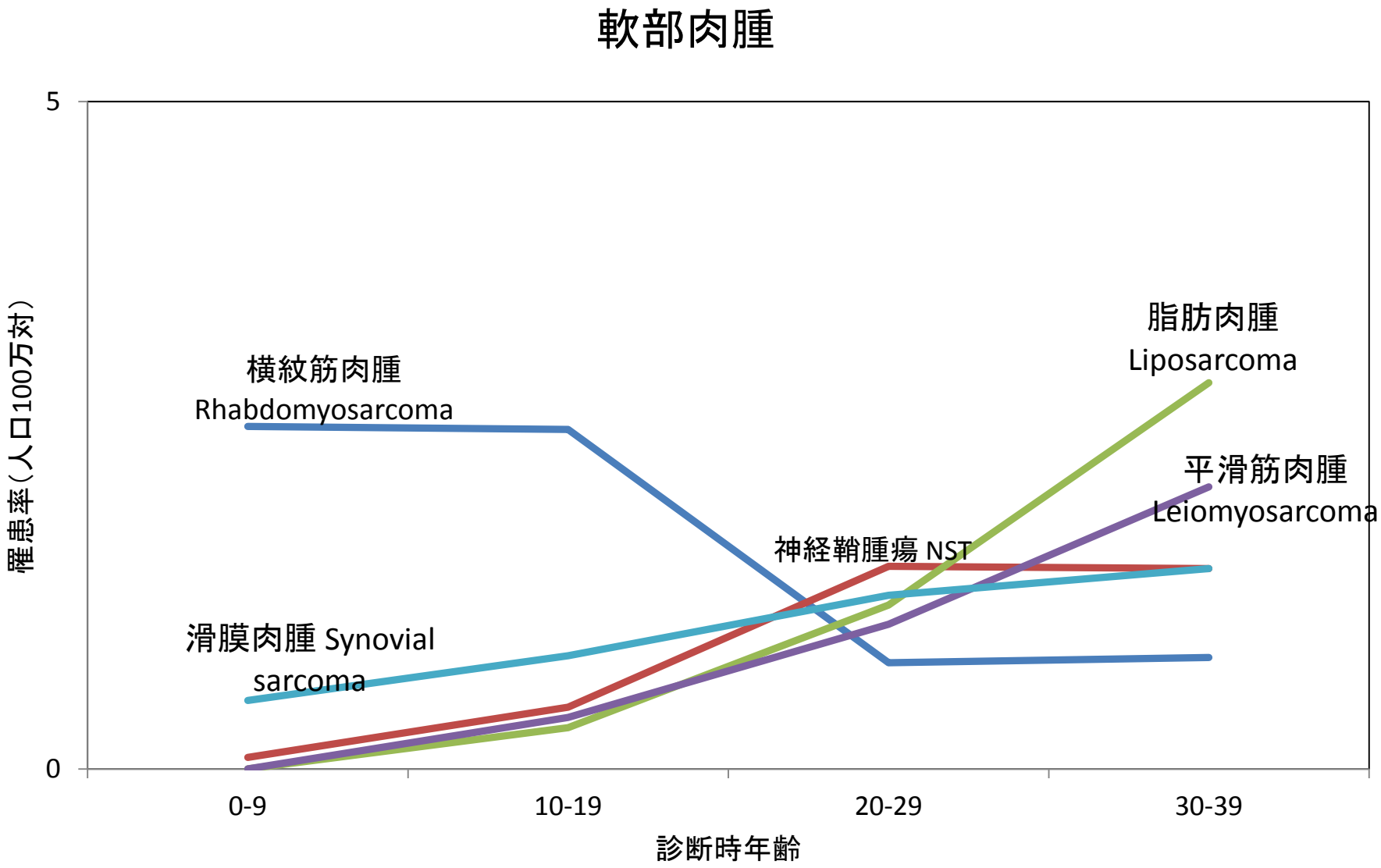

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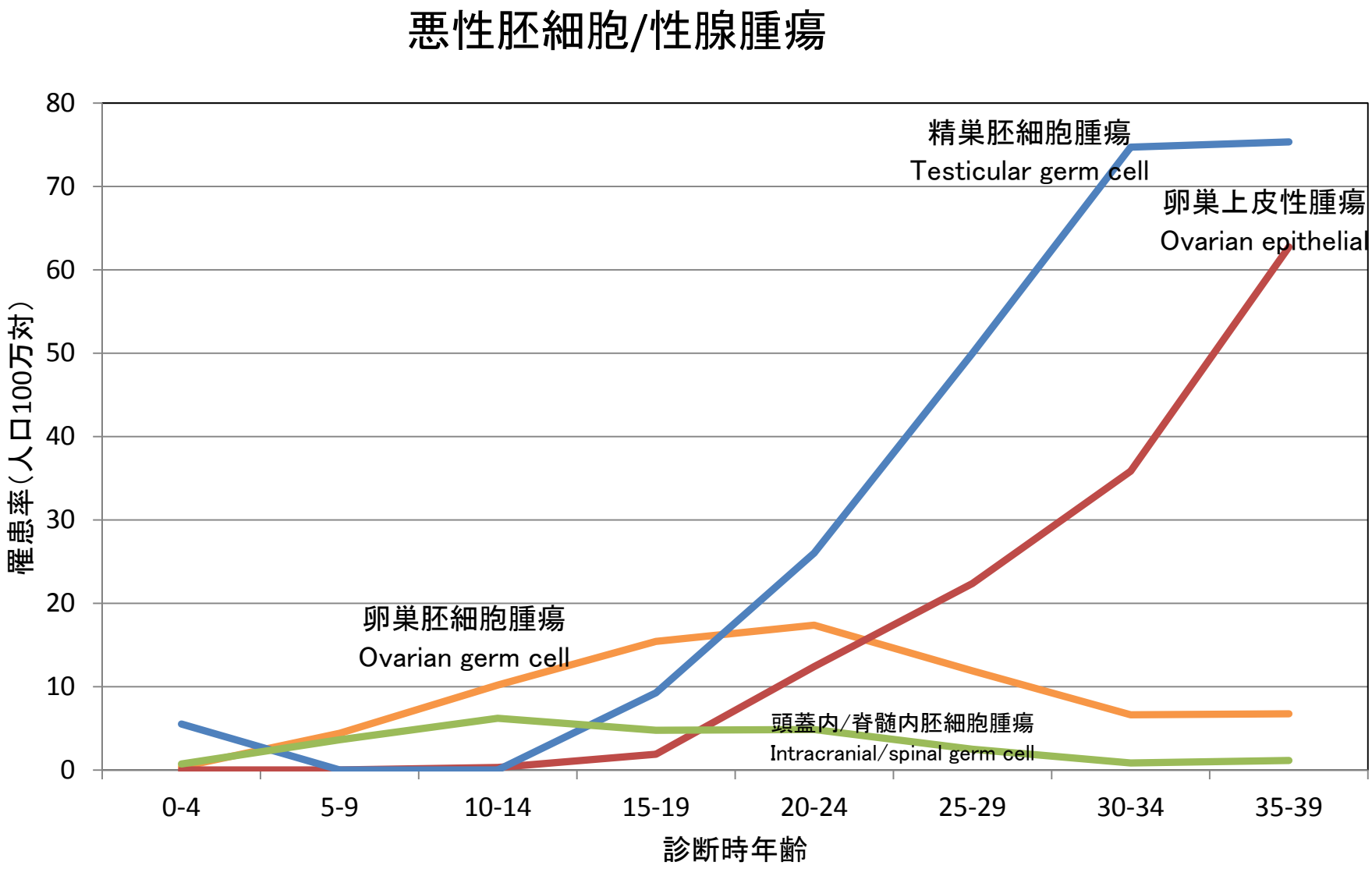

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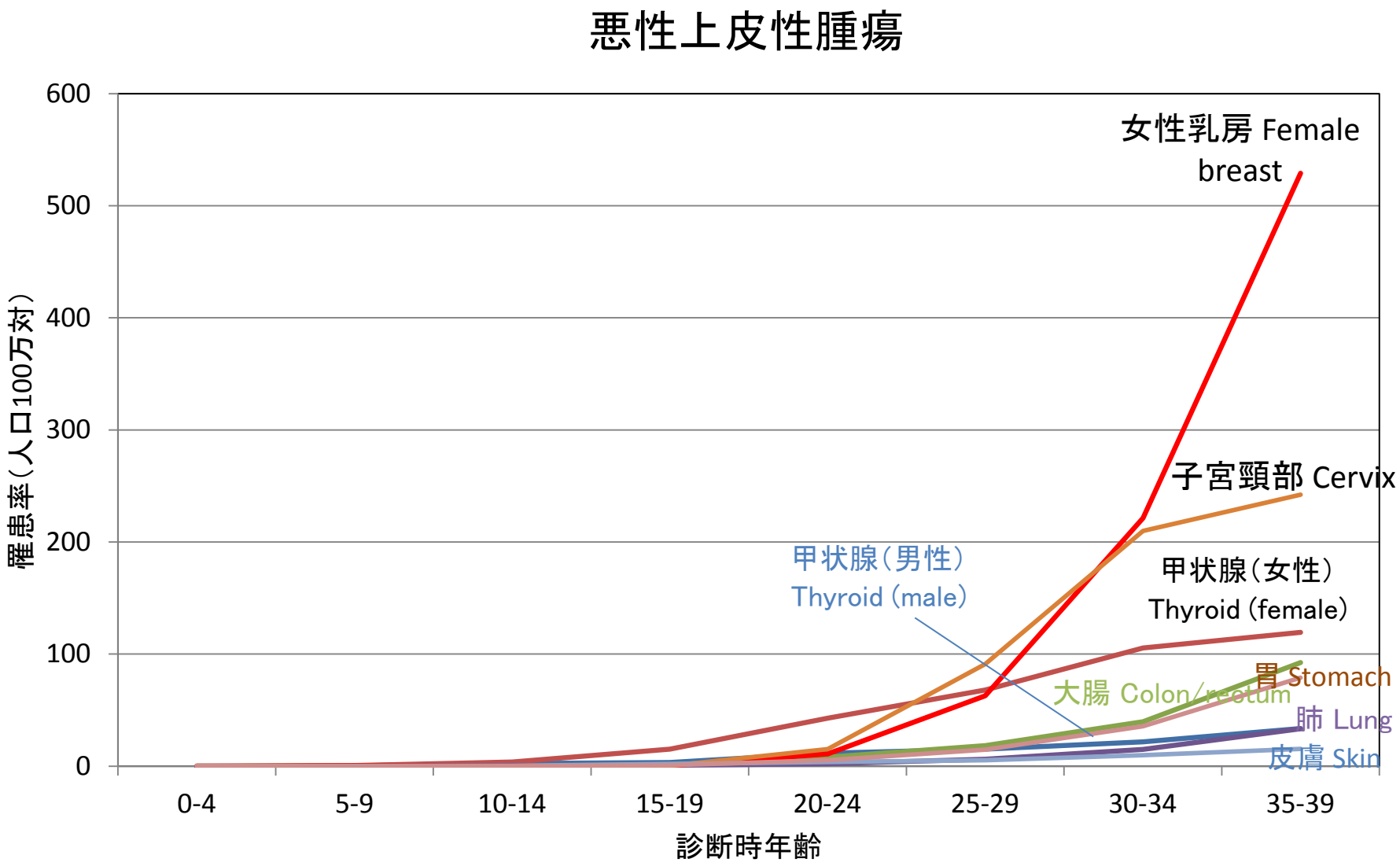

まとめ

- 個々の希少がんは希だが合わせると主要ながんに匹敵する
- 小児からAYA世代にかけてはいわゆる希少がんのほうが多くを占め、その内訳が大きく変化する
- がん登録の精度向上、標準化により、一定の精度で希少がんの統計が算出可能となりつつある
- 全国がん登録データの公表に向けて、治療開発や療養生活に役立つ統計を、より高い精度で、継続性を担保しながら整備してゆく必要がある

ご清聴ありがとうございました