GROUPE SARCOME FRANÇAIS
Les journées annuelles
14es journées annuelles du
GSF-GETO
20, 21 & 22 juin 2018
LE CORUM, PALAIS DES CONGRÈS, MONTPELLIER, FRANCE


Quoi de neuf

Epidémiologie et Santé publique

Le digest …2017-18

Pr S Mathoulin-Pelissier,
Bordeaux, U1219, I Bergonie

- **Tumeurs rares/ Europe**
  - Incidence
  - Survie inégalité
  - Causalité
- **Essais cliniques**

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

Summary

Background Rare cancers pose challenges for diagnosis, treatments, and clinical decision making. Information about rare

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.

	1999–2001 5-year relative survival	2005–07 5-year relative survival	Difference	99·8% CI
Chronic myeloid leukaemia	37·2	57·9	20·7	17·4 to 24·1
Infiltrating duct carcinoma of prostate	67·5	79·8	12·3	6·4 to 18·2
Soft tissue sarcoma of viscera	34·7	43·7	9·0	3·6 to 14·4
Kaposi's sarcoma	75·4	84·2	8·8	1·4 to 16·2
Diffuse B-cell lymphoma	46·9	55·2	8·4	6·5 to 10·2
Follicular B-cell lymphoma	69·5	77·9	8·4	5·9 to 10·8
Poorly differentiated endocrine carcinoma of pancreas and digestive system	25·3	32·7	7·5	2·7 to 12·2
Squamous cell carcinoma with variants of oropharynx	37·5	44·5	7·1	5·0 to 9·2
Soft tissue sarcoma of superficial trunk	43·9	50·4	6·5	1·4 to 11·6
Precursor B/T cell lymphoblastic leukaemia or lymphoma (and Burkitt's leukaemia/lymphoma)	54·3	60·8	6·4	3·8 to 9·1
Plasmacytoma or multiple myeloma (and heavy chain diseases)	29·8	35·0	5·2	3·8 to 6·7
Carcinomas of thyroid gland	85·6	90·6	5·0	3·8 to 6·3
Adenocarcinoma with variants of cervix uteri	63·8	68·8	5·0	1·7 to 8·3
Well differentiated not functioning endocrine carcinoma of pancreas and digestive system	67·7	72·6	4·9	1·5 to 8·4
Soft tissue sarcoma of limbs	63·9	68·4	4·4	1·0 to 7·9

Similarly, the incidence of soft tissue sarcomas was 5 times higher than bone sarcomas, but soft tissue sarcomas were treated centrally less than bone sarcoma.

Cancer Epidemiology 54 (2018) 95–100

Contents lists available at ScienceDirect

Cancer Epidemiology

journal homepage: www.elsevier.com/locate/canep

Bayesian estimates of the incidence of rare cancers in Europe

Laura Botta[a,*], Riccardo Capocaccia[a], Annalisa Trama[a], Christian Herrmann[b,c,d], Diego Salmerón[e,f,g], Roberta De Angelis[h], Sandra Mallone[h], Ettore Bidoli[i], Rafael Marcos-Gragera[j], Dorota Dudek-Godeau[k], Gemma Gatta[a], Ramon Cleries[l,m], The RARECAREnet Working group[1]

Moins de 150 cas/ ans : un intérêt sinon méthode habituelle

Epidemiology of rare cancers and inequalities in oncologic outcomes

G. Gatta*, A. Trama, R. Capocaccia, RARECARENet Working Group

Evaluative Epidemiology Unit, Fondazione IRCCS, Istituto Nazionale dei Tumori, Milan, Italy

Table 4
Number of cases (No.) and 5-year Relative Survival (RS%) with (95% CI) for selected rare cancers by age.

Family	Rare cancer entity	age class							
		0–14		15–24		25–64		65+	
		No.	5-year RS	No.	5-year RS	No.	5-year RS	No.	5-year RS
soft tissue sarcomas	Soft tissue sarcoma (STS)	2222	68 (66–71)	2631	64 (62–66)	37,226	64 (63–64)	30,625	46 (45–47)
	STS of head and neck	174	71 (65–79)	128	64 (55–74)	1553	67 (64–70)	2207	53 (50–56)
	STS of limbs	257	87 (83–92)	651	78 (74–81)	8541	74 (73–75)	7645	58 (57–60)
	STS of superficial trunk	145	59 (51–68)	234	59 (53–66)	3775	57 (55–59)	3569	37 (35–39)
	STS of mediastinum	11	63 (40–100)	20	23 (8–63)	262	27 (22–33)	164	15 (9–23)
	STS of heart	5	40 (14–100)	14	(NE)	143	15 (10–24)	41	6 (2–23)
	STS of breast	2	50 (13–100)	60	85 (76–96)	1794	81 (79–83)	1008	61 (57–66)
	STS of uterus	1	100 (NE)	38	78 (66–93)	5829	59 (58–60)	2700	36 (34–38)
	Other STSs of genitourinary tract	132	60 (52–70)	97	71 (62–81)	1447	60 (57–63)	1431	38 (34–41)
	STS of viscera	65	66 (55–79)	69	53 (42–67)	2587	45 (43–47)	3194	39 (37–41)
	STS of paratestis	5	100 (NE)	15	52 (30–91)	222	92 (87–97)	268	85 (77–94)
	STS of retroperitoneum and peritoneum	11	73 (51–100)	48	42 (28–61)	2444	46 (44–48)	2351	31 (28–33)
	STS of pelvis	63	62 (50–76)	93	53 (44–65)	1569	55 (53–58)	1339	36 (33–40)
	STS of skin	67	98 (94–100)	258	99 (97–100)	2731	96 (95–97)	1672	77 (73–81)
	STS of paraorbit	34	74 (59–92)	11	46 (22–96)	32	65 (49–88)	38	57 (37–88)
	STS of brain and other nervous system	147	49 (40–58)	209	59 (52–66)	1573	59 (56–62)	766	44 (40–49)
	Embryonal rhabdomyosarcoma of soft tissue	584	76 (72–79)	148	48 (40–58)	80	40 (29–53)	13	11 (2–75)
	Alveolar rhabdomyosarcoma of soft tissue	238	49 (43–57)	128	24 (17–34)	117	24 (17–34)	32	29 (14–61)
	Ewin's sarcoma of soft tissue	167	67 (60–76)	270	51 (45–58)	423	40 (35–45)	132	21 (14–31)

Epidemiology of rare cancers and inequalities in oncologic outcomes

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		No.	5-year RS	No.	5-year RS	No.	5-year RS	No.	5-year RS
soft tissue sarcomas	Soft tissue sarc						63–64)	30,625	46 (45–47)
	STS of head an						64–70)	2207	53 (50–56)
	STS of limbs						73–75)	7645	58 (57–60)
	STS of superfic						55–59)	3569	37 (35–39)
	STS of mediast						22–33)	164	15 (9–23)
	STS of heart						10–24)	41	6 (2–23)
	STS of breast						79–83)	1008	61 (57–66)
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Age affects survival and about 50% of all rare cancers occurs in the elderly (65+) [5]. This was relevant for some rare cancers chosen to be described by this monograph. Actually, 50% or more cases occurred in the elderly for the epithelial tumours of nasal cavities, STS of H&N, viscera and paratestis. This point makes difficult to plan clinical trials, in which elderly are not usually included, and finally the question of disparities in accessing to proper treatment remains quite unsolved in the elderly.

Parikh *et al. Clin Sarcoma Res (2018) 8:8*
https://doi.org/10.1186/s13569-018-0094-x

Clinical Sarcoma Research

RESEARCH Open Access

Treatment patterns and survival among older adults in the United States with advanced soft-tissue sarcomas

Rohan C. Parikh[1*], Maria Lorenzo[2], Lisa M. Hess[3], Sean D. Candrilli[1], Steven Nicol[3] and James A. Kaye[4]

Vineis *et al. Emerg Themes Epidemiol (2017) 14:7*
DOI 10.1186/s12982-017-0061-7

Emerging Themes in Epidemiology

COMMENTARY Open Access

Causality in cancer research: a journey through models in molecular epidemiology and their philosophical interpretation

Paolo Vineis[1*†], Phyllis Illari[2†] and Federica Russo[3†]

- **Tumeurs rares/ Europe**
- **Essais cliniques**
  - Design pour des maladies rares
  - Revues systématiques et Sarcomes
  - Les patients agées et sarcomes

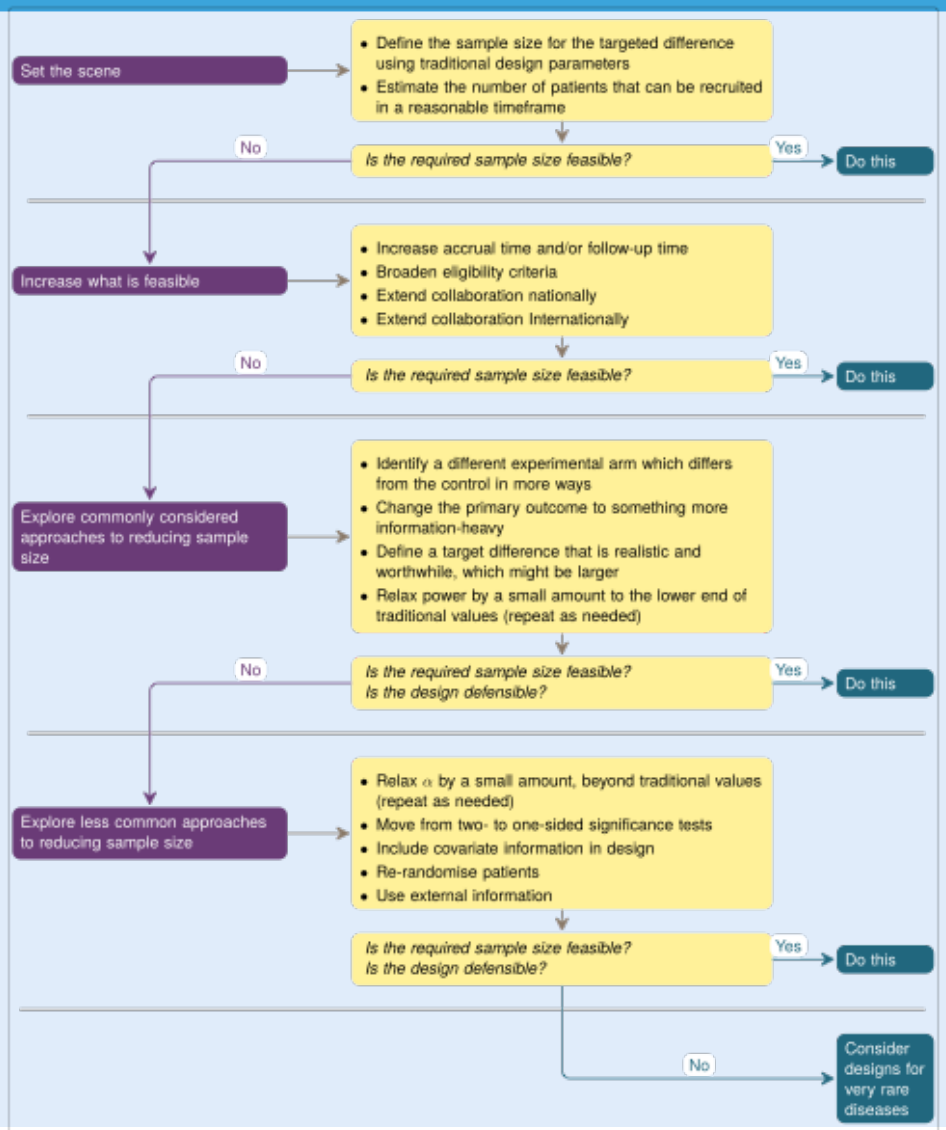
Parmar *et al. BMC Medicine* (2016) 14:183
DOI 10.1186/s12916-016-0722-3

BMC Medicine

CORRESPONDENCE Open Access

How do you design randomised trials for smaller populations? A framework

Mahesh K. B. Parmar[1], Matthew R. Sydes[1] and Tim P. Morris[1,2*]



Plus de temps sur la réflexions des designs+++

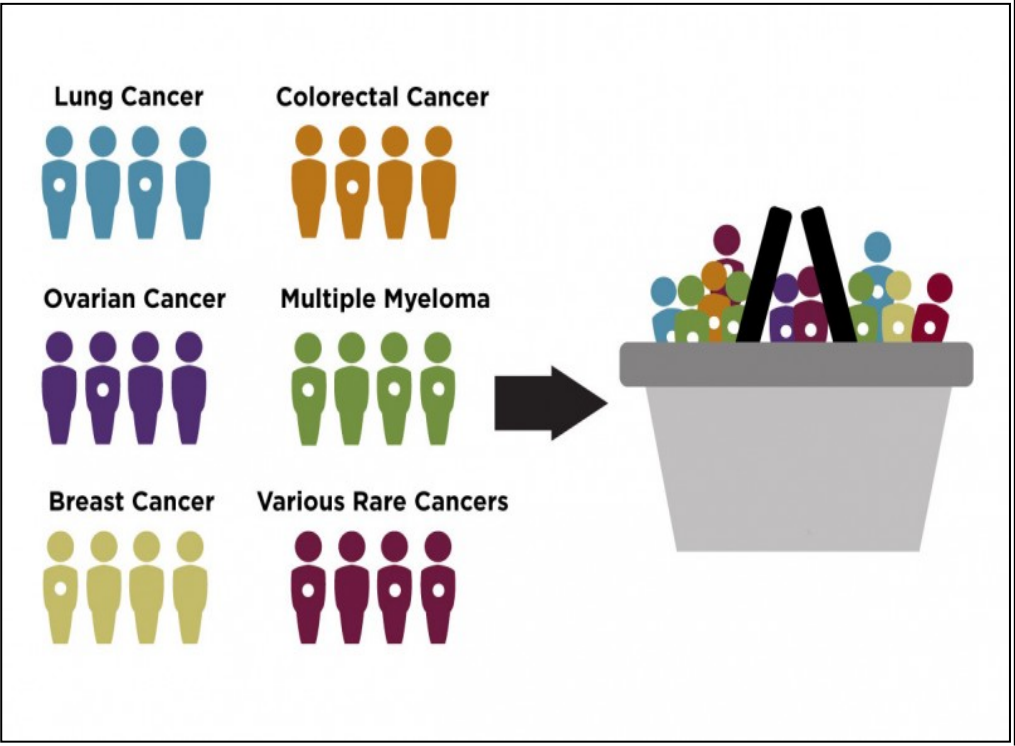
Qq pistes

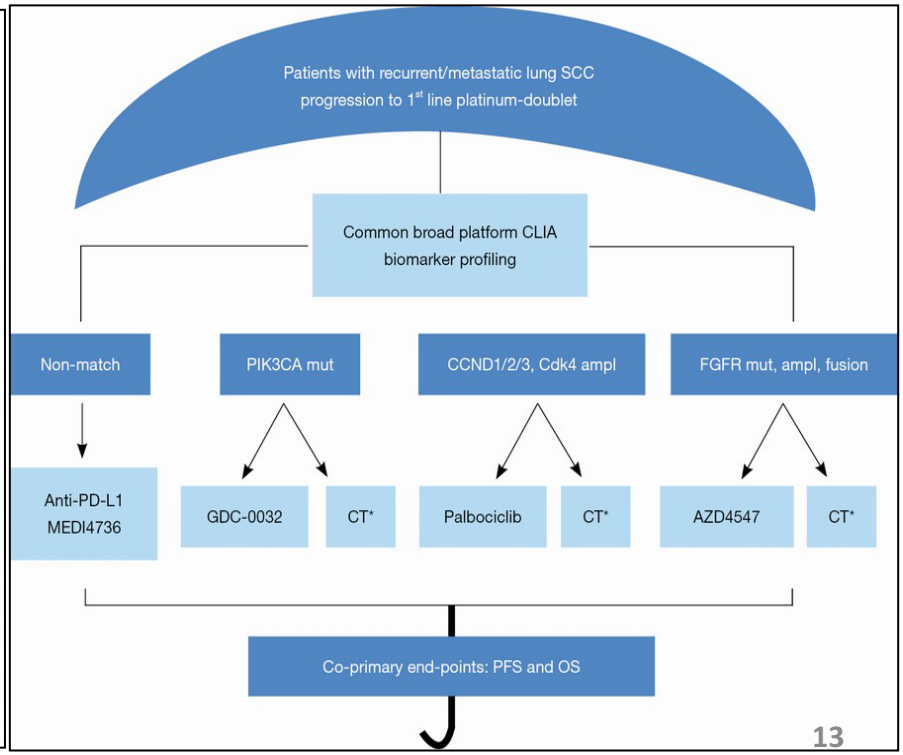
Exemple : Euramos 1 trial

Next Generation Sequencing (NGS) → Plus qu'un biomarqueur → plusieurs molécules peuvent être évaluées dans un essai clinique

L. A. Renfro, D.J Sargent, revue 2017

- **Master Protocol**
- **Basket – 1 altération ciblée**
- **Umbrella - 1 Tumeur**





Cancer Treatment Reviews 63 (2018) 71–78

Contents lists available at ScienceDirect

Cancer Treatment Reviews

journal homepage: www.elsevierhealth.com/journals/ctrv

Systematic or Meta-analysis Studies

Multi-agent chemotherapy in advanced soft tissue sarcoma (STS) – A systematic review and meta-analysis

Alona Zer [a,b,*], Rebecca M. Prince [c,d], Eitan Amir [c,d,e], Albiruni R. Abdul Razak [c,d]

[a] Rabin Medical Center, Petach Tikva, Israel
[b] Faculty of Medicine, Tel Aviv University, Israel
[c] Princess Margaret Cancer Centre, Toronto, Canada
[d] Department of Medicine, University of Toronto, Toronto, Canada
[e] Institute of Health Policy Management and Evaluation, University of Toronto, Toronto, Canada

Conclusion: Multi-agent chemotherapy is associated with a modest, but statistically significant improvement in outcomes in STS. Combining chemotherapy with non-cytotoxic agents might represent a promising strategy.

The Oncologist®

Sarcomas

Outcomes of Elderly Patients with Advanced Soft Tissue Sarcoma Treated with First-Line Chemotherapy: A Pooled Analysis of 12 EORTC Soft Tissue and Bone Sarcoma Group Trials

EUGENIE YOUNGER [a], [a]
ROBIN LEWIS JONES, [a] AL

Background. Almost half of patients diagnosed with soft tissue sarcoma (STS) are older than 65 years; however, the outcomes of elderly patients with metastatic disease are not well described.

Patients and Methods. An elderly cohort of patients aged ≥65 years was extracted from the European Organization for Research and Treatment of Cancer (EORTC) Soft Tissue and Bone Sarcoma Group database of patients treated with first-line chemotherapy for advanced STS within 12 EORTC clinical trials. Endpoints were overall survival (OS), progression-free survival (PFS), and response rate (RR).

Results. Of 2,810 participants in EORTC trials, there were 348 elderly patients (12.4%, median 68 years; interquartile range [IQR], 67–70; maximum 84 years) and 2,462 patients aged <65 years (median 49 years; IQR, 39–57). Most elderly patients had a performance status of 0 ($n = 134$; 39%) or 1 ($n = 177$; 51%). Leiomyosarcoma ($n = 130$; 37%) was the most common histological subtype. Lung metastases were present in 181 patients (52%) and liver metastases in 63 patients (18%). Overall, 126 patients (36%) received doxorubicin, 114 patients (33%) doxorubicin + ifosfamide, 43 patients (12%) epirubicin, 39 patients (11%) trabectedin, and 26 patients (7%) ifosfamide. Overall RR was 14.9% ($n = 52$), median PFS was 3.5 months (95% confidence interval [CI], 2.7–4.3), and median OS was 10.8 months (95% CI, 9.43–11.83). In patients aged <65 years, overall RR was 20.3% ($n = 501$), median OS was 12.3 months (95% CI, 11.9–12.9), and median PFS was 4.3 months (95% CI, 3.9–4.6).

Conclusion. Elderly patients with metastatic STS treated with first-line chemotherapy were largely underrepresented in these EORTC STS trials. Their outcomes were only slightly worse than those of younger patients. Novel trials with broader eligibility criteria are needed for elderly patients. These trials should incorporate geriatric assessments and measurements of age-adjusted health-related quality of life.

The Oncologist 2018;23:1–10

Le G8 pour l'intervention/ détection et le G-Code pour les données dans les essais cliniques *(article soumis en revision): PACAN-Dialog-Sofog*

Development of the initial geriatric core dataset by 7 pairs of geriatricians (Committee 1, part 1)
January-October 2015

↓

Modified Delphi consensus process by three rounds (Committee 1, part 2)
July-September 2016

↓

Assessment of face validity about the final geriatric core data set by 2 large panels of health professionals: national (55, Committee 2) and international panels (42, Committee 3)
March-September 2017

Supervision by a Steering Committee:
Consensus approach, identification and contact with experts, development of questionnaires and analyses of results

Final G-CODE with 10 tools[b]	**ADL and 4-IADL** **Mini-Cog** **Mini-GDS** **UpCCI** **BMI and weight loss** **TUG** **"Do you live alone?" Y/N** **"Do you have a person or caregiver to help you?" Y/N**

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Merci de votre attention

Plateforme de recherche clinique

PACAN

Personnes Âgées et CANcer

LA LIGUE CONTRE LE CANCER