ESMO SARCOMA & GIST

DESMOPLASTIC SMALL ROUND CELL TUMORS

JY Blay

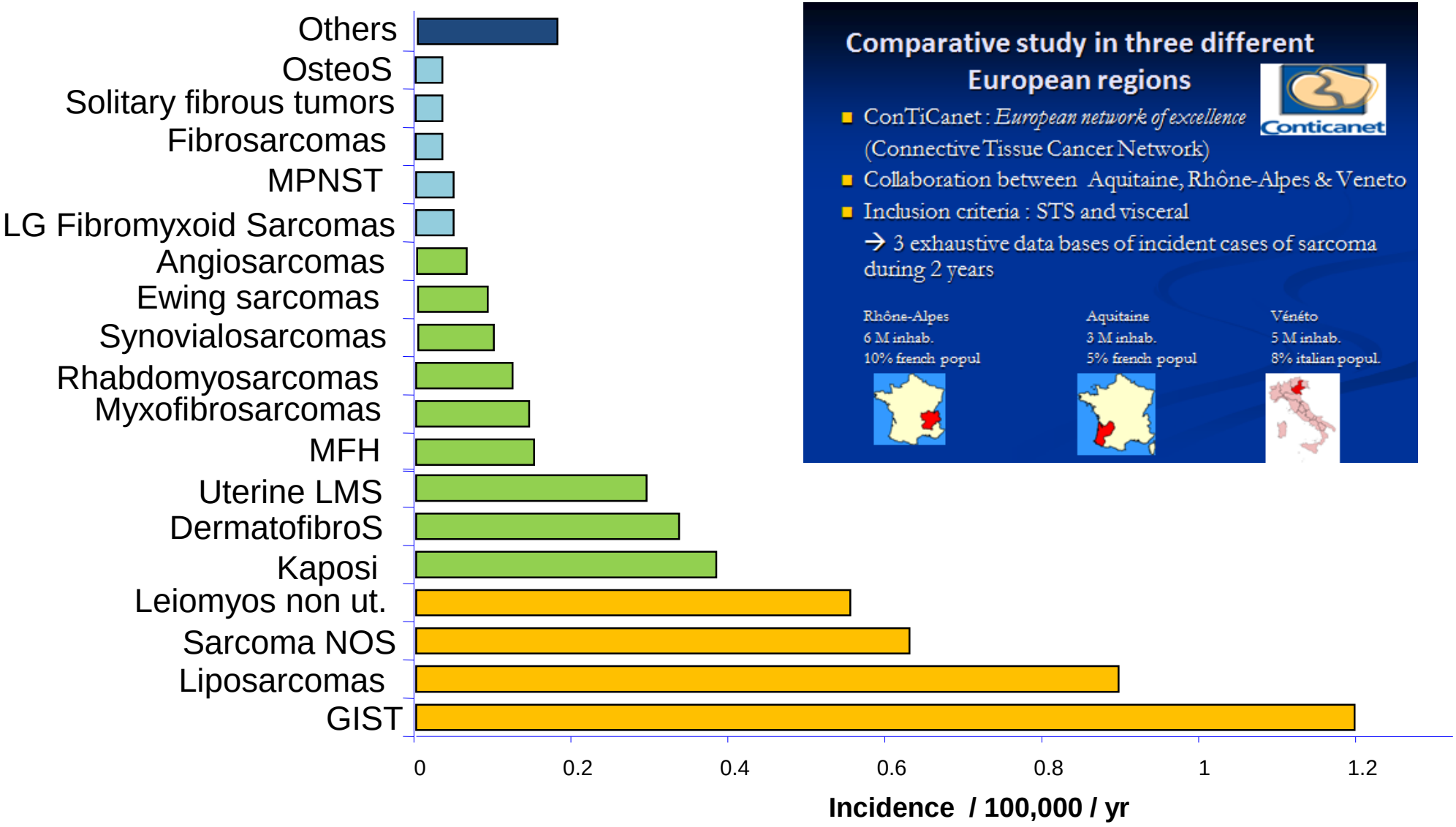
DISCLOSURE SLIDE

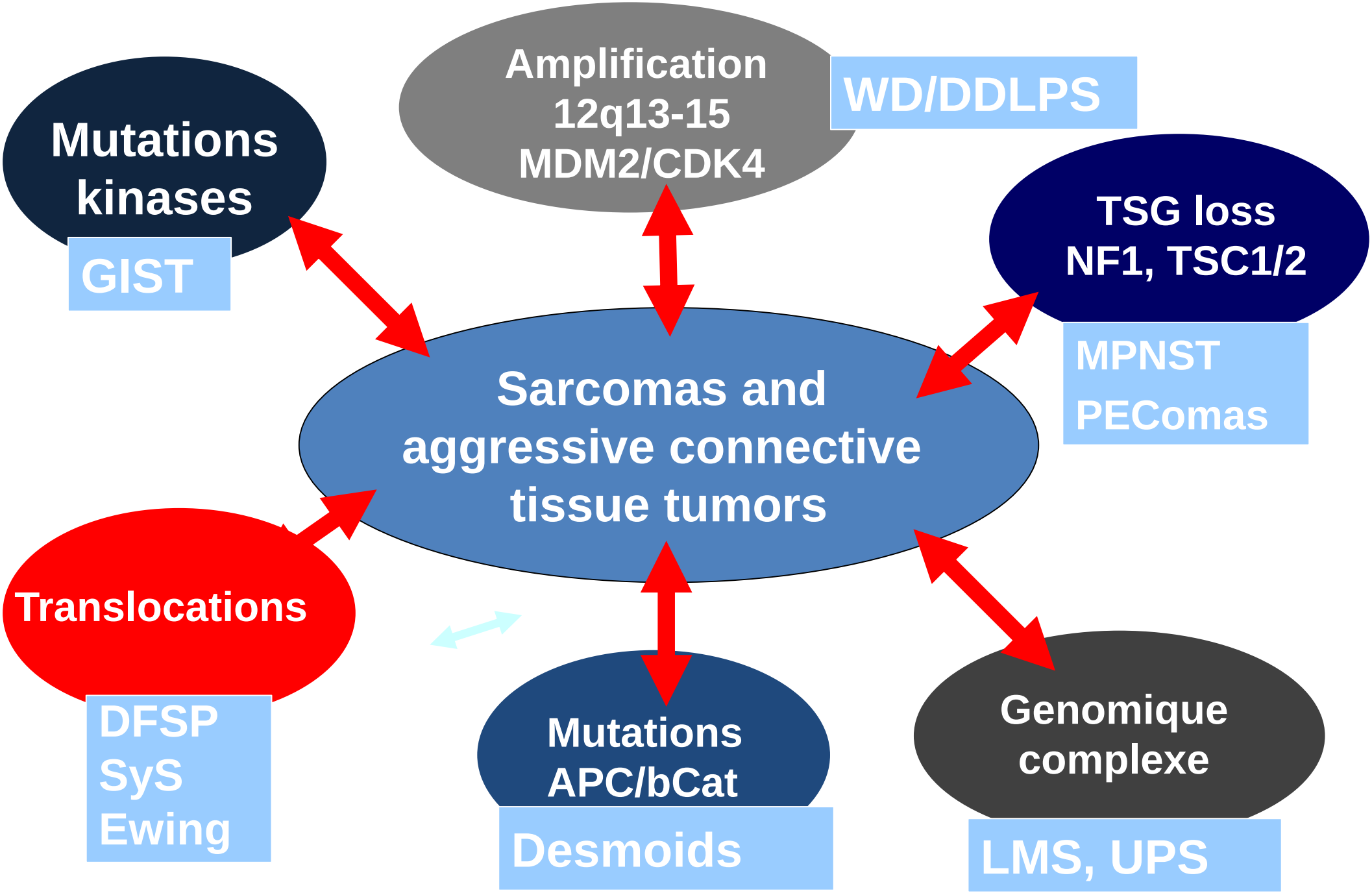
Company	Scientific advice	Scientific works	Symposia & oral communication
Abbvie	X	X	
Amgen	X	X	X
ARIAD	X	X	
AstraZeneca		X	X
Bayer	X	X	X
BMS	X	X	X
Deciphera	x	x	
DDB	X	X	
EISAI	X	X	X
Genomic Health		X	X
Gilead		X	X
GSK		X	X
INNATE PHARMA	X (member of the Supervisory committee)		
INCYTE		X	
IQVIA	x	x	x
Jansenn		X	X
LILLY		X	X
Merck Serono		X	X
MSD		X	X
Nanobiotix	X	x	
Novartis	X	X	X
Novex		X	X
Onxeo	X		
Pfizer		X	X
Pharmamar	X	X	
PRA		X	
Roche		X	X
Sanofi Aventis		X	X
Swedish Orphan		X	X
Takeda		X	
Toray		X	

Topics

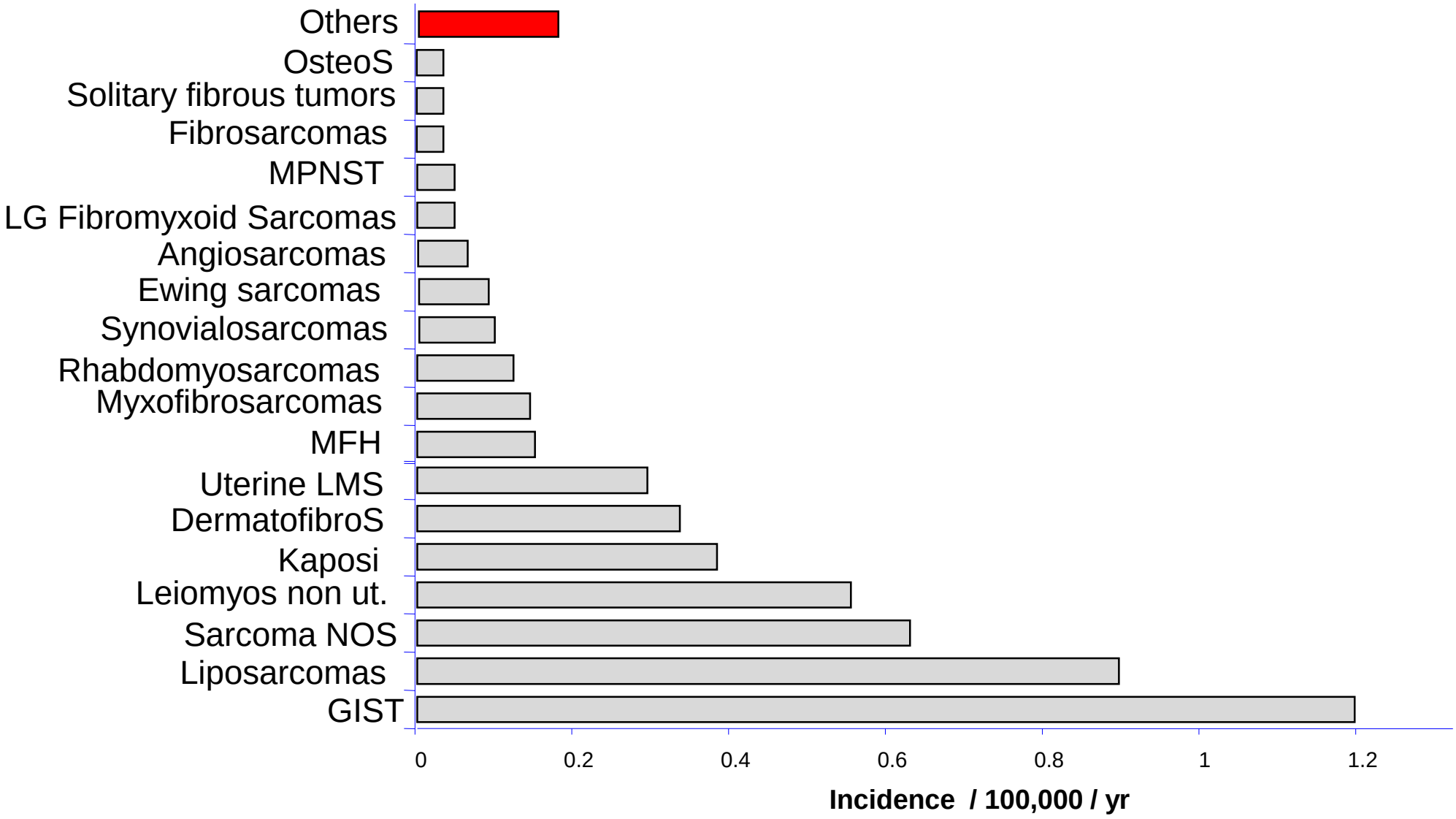
- DSCRT
- General guidelines
- Local treatments, surgery, HIPEC, WART…
- Systemic treatments

Over 145 histotypes of sarcomas…




Amplification
12q13-15
MDM2/CDK4
WD/DDLPS
Mutations
kinases
GIST
TSG loss
NF1, TSC1/2
MPNST
PEComas
Sarcomas and
aggressive connective
tissue tumors
Translocations
DFSP
SyS
Ewing
Mutations
APC/bCat
Desmoids
Genomique
complexe
LMS, UPS

Incidence of sarcoma



Management of Desmoplastic Small Round-cell Tumors in Children and Young Adults

Andrea Hayes Jordan, MD and Alberto Pappo, MD†*

(*J Pediatr Hematol Oncol* 2012;34:S73–S75)

Described in 1989
t(11,22), EWS-WT1
Clinical presentation

- Age 5-25 , until 63 ans
- Man 90%
- Advanced (unless incidental discovery)

0,17/10^{e}6 /year in France
13 per year

Abdomen+++ (rare other sites)

Peritoneal, liver, lung, bone metastasesosseuses

Table 1: Patient and tumor characteristics			
Number of patients			48 (100%)
Median age, years [range]			22 [3 - 57]
Gender			
	Male		35 (73%)
	Female		13 (27%)
WHO performance status			
	0		28 (58%)
	1		6 (13%)
	2		1 (2%)
	N/A		13 (27%)
Median PCI [range]			9 [2 - 27]
Lymph node metastases			
	Yes		14 (29%)
	No		34 (71%)
MD Anderson stage			
	I		21 (44%)
	II		10 (21%)
	N/A		17 (35%)
Abbreviations: WHO, World Health Organization;			
PCI, peritoneal cancer index; N/A, non-available			

C. Honoré et al, 2017

Management of Desmoplastic Small Round-cell Tumors in Children and Young Adults

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(*J Pediatr Hematol Oncol* 2012;34:S73–S75)

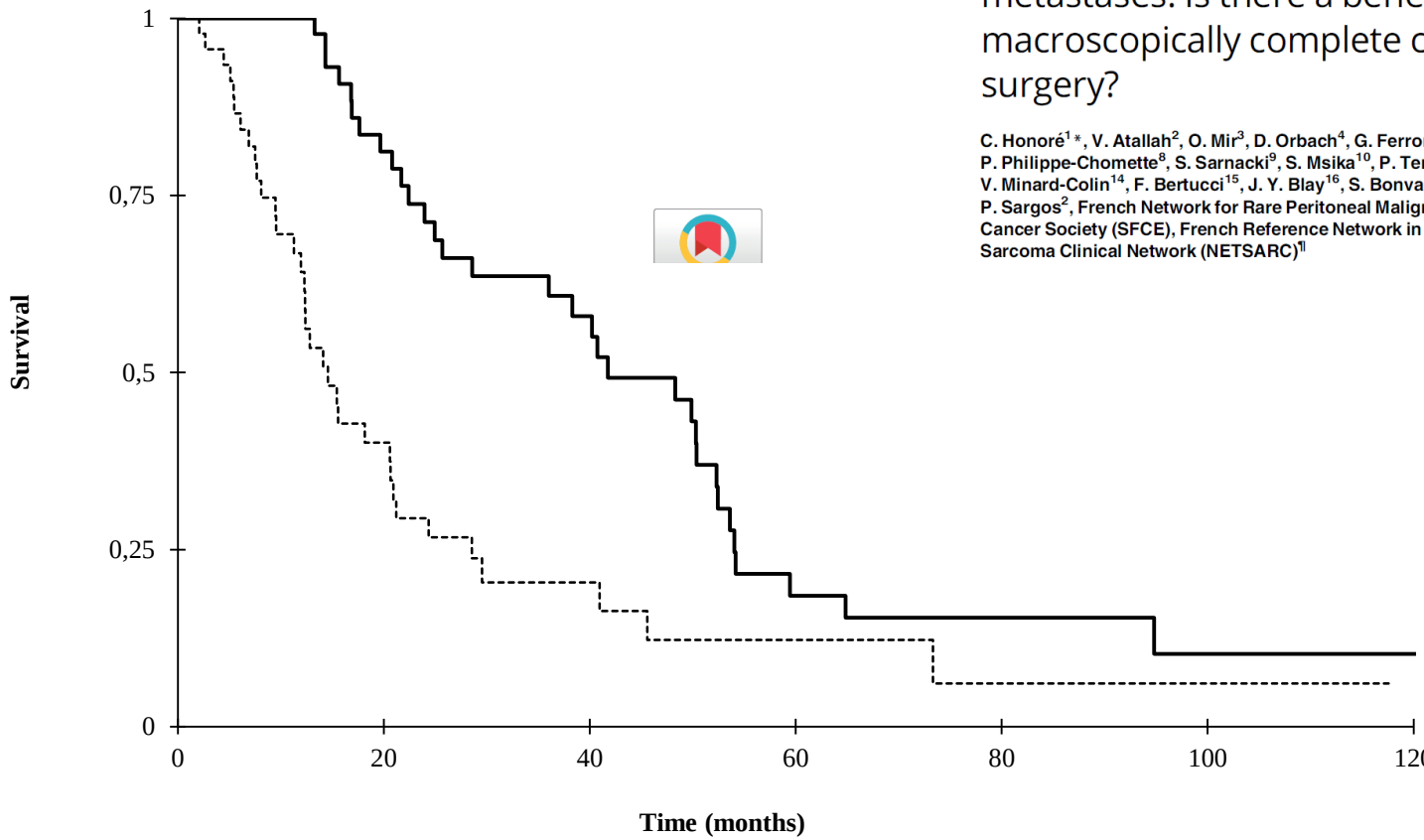
Chemotherapy regimen similar to those of Ewing (?)

Cytoreductive surgery

WA-Radiotherapy (?)

HIPEC?

PLOS ONE



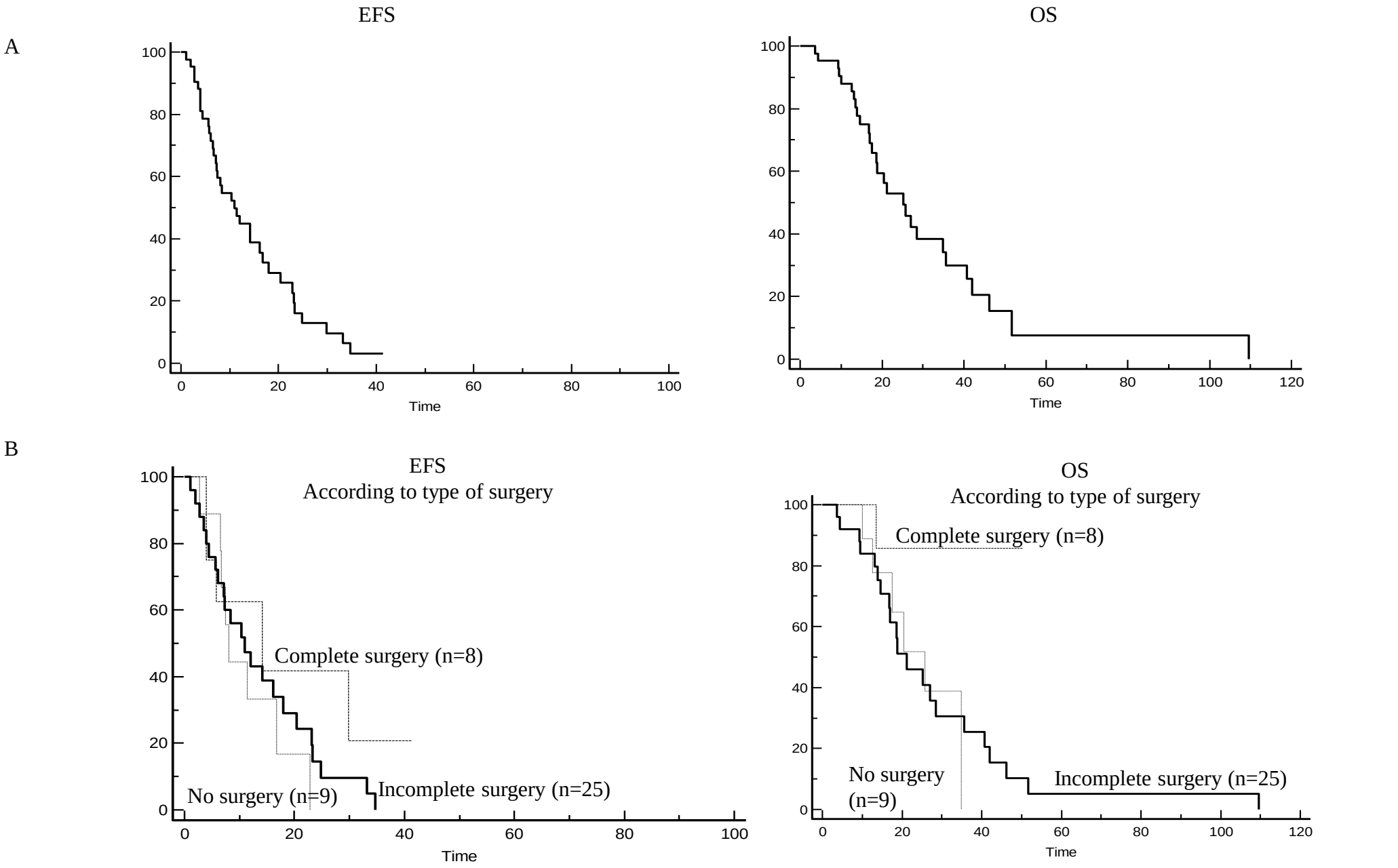
RESEARCH ARTICLE

Abdominal desmoplastic small round cell tumor without extraperitoneal metastases: Is there a benefit for HIPEC after macroscopically complete cytoreductive surgery?

C. Honoré[1]*, V. Atallah[2], O. Mir[3], D. Orbach[4], G. Ferron[5], C. LePéchoux[6], J. B. Delhorme[7], P. Philippe-Chomette[8], S. Sarnacki[9], S. Msika[10], P. Terrier[11], O. Glehen[12], H. Martelli[13], V. Minard-Colin[14], F. Bertucci[15], J. Y. Blay[16], S. Bonvalot[17], D. Elias[1], A. LeCesne[3], P. Sargos[2], French Network for Rare Peritoneal Malignancies (RENAPE), French Pediatric Cancer Society (SFCE), French Reference Network in Sarcoma Pathology (RRePS) French Sarcoma Clinical Network (NETSARC)[¶]

C. Honoré et al, 2017

Figure 3


A
EFS
OS
100
80
60
40
20
0
0
20
40
60
80
100
120
Time
B
EFS
According to type of surgery
Complete surgery (n=8)
No surgery (n=9)
Incomplete surgery (n=25)
OS
According to type of surgery
Complete surgery (n=8)
No surgery
(n=9)
Incomplete surgery (n=25)

Published OnlineFirst June 5, 2018; DOI: 10.1158/1078-0432.CCR-18-0202

Translational Cancer Mechanisms and Therapy

Clinical Cancer Research

Multimodality Treatment of Desmoplastic Small Round Cell Tumor: Chemotherapy and Complete Cytoreductive Surgery Improve Patient Survival

Vivek Subbiah[1], Salah-Eddine Lamhamedi-Cherradi[1], Branko Cuglievan[2], Brian A. Menegaz[1], Pamela Camacho[2], Winston Huh[2], Vandhana Ramamoorthy[1], Pete M. Anderson[3], Raphael E. Pollock[4], Dina C. Lev[4], Wei Qiao[5], Mary Frances McAleer[6], Robert S. Benjamin[1], Shreyaskumar Patel[1], Cynthia E. Herzog[2], Najat C. Daw[2], Barry W. Feig[7], Alexander J. Lazar[8], Andrea Hayes-Jordan[7], and Joseph A. Ludwig[1]

Table 1. Desmoplastic small round cell tumor clinical trials

	Cancer Center				
Published clinical trials[b]	**This Study**	**MSKCC**	**MDACC**	**UK Centers**	**France GR Center**
Study dates	1990–2016	1972–2003	Until 1998	1991–2012	1991–2013
Patient demographics					
No. of patients	187	66	39	41	38
Median age, y	22.6	19	25	27	27
Male:female ratio	4.8:1	10:1	4.5:1	3.1:1	3.5:1
Caucasian race	75.4%	75%	N/A	N/A	N/A
Treatment modality (% of patients)					
Chemotherapy	98%	>92%	92%	93%	100%
Surgery	61%	71%	92%	20%	60.5%
Radiotherapy	49%	>44%	N/A	15%	30%
HIPEC[a]	72%	N/A	N/A	N/A	5%
SCT	6.4%	0%	0%	0%	N/A
Survival					
Median survival (mo)	35	28	N/A	16	25.7
3-y OS rate	48.2%	44%	N/A	N/A	31.6%
5-y OS rate	21.6%	15%	N/A	N/A	8.3%
Improved by surgery	Yes ($P = 0.04$)	Yes		Yes ($P = 0.024$)	Yes ($P = 0.026$)
Improved by radiation	Yes ($P < 0.01$)	Yes		Yes ($P = 0.015$)	Yes ($P = 0.022$)
Improved by HIPEC	No ($P = 0.26$)	N/A	N/A	N/A	N/A

Abbreviation: GR Center, Gustave Roussy.

[a]HIPEC was only offered to patients that had undergone a CCS ($n = 114$).

[b]More limited studies include earlier ones conducted at MSKCC (40) and MDACC (41, 42), as well as contemporary studies performed in the UK (20) and France (8).

Topics

- DSCRT
- General guidelines
- Local treatments, surgery, HIPEC, WART…
- Systemic treatments

clinical practice guidelines

Annals of Oncology 23 (Supplement 7): vii92–vii99, 2012
doi:10.1093/annonc/mds253

Soft tissue and visceral sarcomas: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up†

The ESMO / European Sarcoma Network Working Group*

clinical practice guidelines

Annals of Oncology 23 (Supplement 7): vii92–vii99, 2012
doi:10.1093/annonc/mds253

Soft tissue and visceral sarcomas: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up†

The ESMO / European Sarcoma Network Working Group*

- Biopsy first
  - Assessment by an experienced team
- En bloc surgical resection
  - Planning R0
  - If R1, consider re resection
- Post operative radiotherapy
  - (G2-3 and/or deep seated, and/or >5cm)
- Preoperative radiotherapy

NetSARC: a network of 26 sarcoma reference centers in France
35784 pts with follow-up presented in MDT since 2010

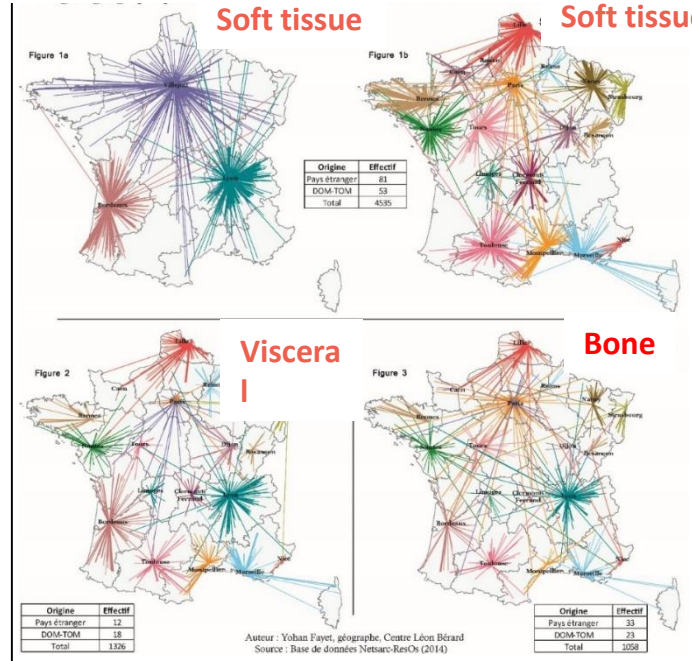
- 26 centers of reference in **Netsarc**
- Linked with Pathology network (**RREPS**)
- Linked with Bone Network **RESOS** (2014)

- 3 networks to be merged (2019)

- Single website

- Entry in the site by CRAs
- Not a clinical trial, a registry

- **Aims:**
  - Guidelines
  - Guiding best practices/patient pathways
  - Measuring
  - Research



Websites

- netsarc.org
- rreps.org
- resos.org

MUNICH 2018 ESMO congress

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ESMO

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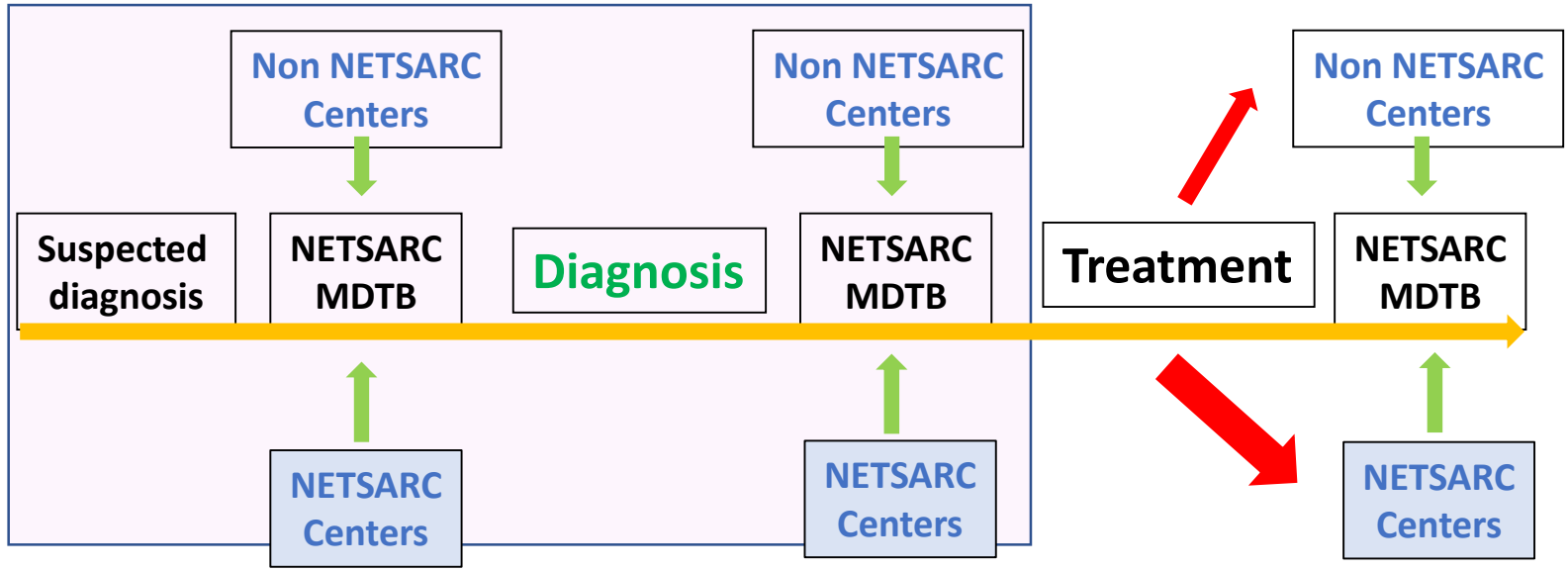
doi:10.1093/annonc/mdx484
Published online 28 August 2017

ORIGINAL ARTICLE

Improved survival using specialized multidisciplinary board in sarcoma patients

J.-Y. Blay[1,2*], P. Soibinet[3], N. Penel[4], E. Bompas[5], F. Duffaud[6], E. Stoeckle[7], O. Mir[8], J. Adam[8], C. Chevreau[9], S. Bonvalot[8,10], M. Rios[11], P. Kerbrat[12], D. Cupissol[13], P. Anract[14], F. Gouin[15], J.-E. Kurtz[16], C. Lebbe[17], N. Isambert[18], F. Bertucci[19], M. Toumonde[7], A. Thyss[20], S. Piperno-Neumann[10], P. Dubray-Longeras[21], P. Meeus[1,2], F. Ducimetière[1,2], A. Giraud[7], J.-M. Coindre[7], I. Ray-Coquard[1,2], A. Italiano[7†] & A. Le Cesne[8†], on behalf of the NETSARC/RREPS and French Sarcoma Group–Groupe d'Etude des Tumeurs Osseuses (GSF-GETO) networks[‡]

- Does presentation of the patient to a NetSARC MDT prior to treatment impact on management and prognosis?



Results

MDT before treatment

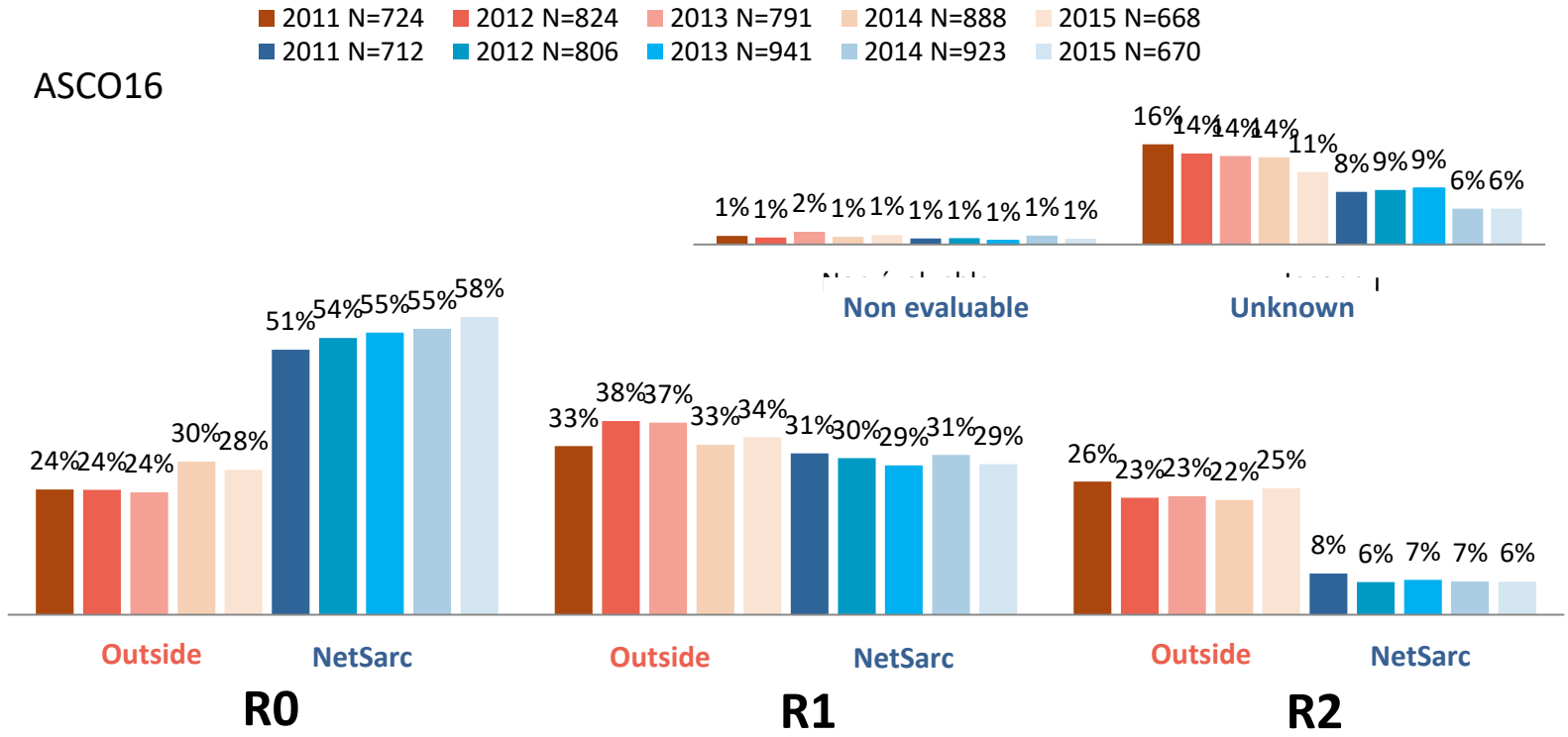
- Overall 37% were presented to a Netsarc multidisciplinary board (NMTB) prior to initial treatment
- Between 2010 and 2015, the proportion of pts reviewed in Netsarc MDT prior to surgery increased from 30,3% to 41,6% .

Results (3)

Better management when MDT before treatment

- A higher number of pts presented in Netsarc MDTB had
  - Adequate imaging of the tumor before treatment/ surgery (87,9% vs 67,8%, $p<0.0001$)
  - Biopsy prior the first resection (87,% vs 55,0%, $p<0.0001$).

Quality of initial surgery, incident patients (STS & visceral sarcomas operated)



ESMO

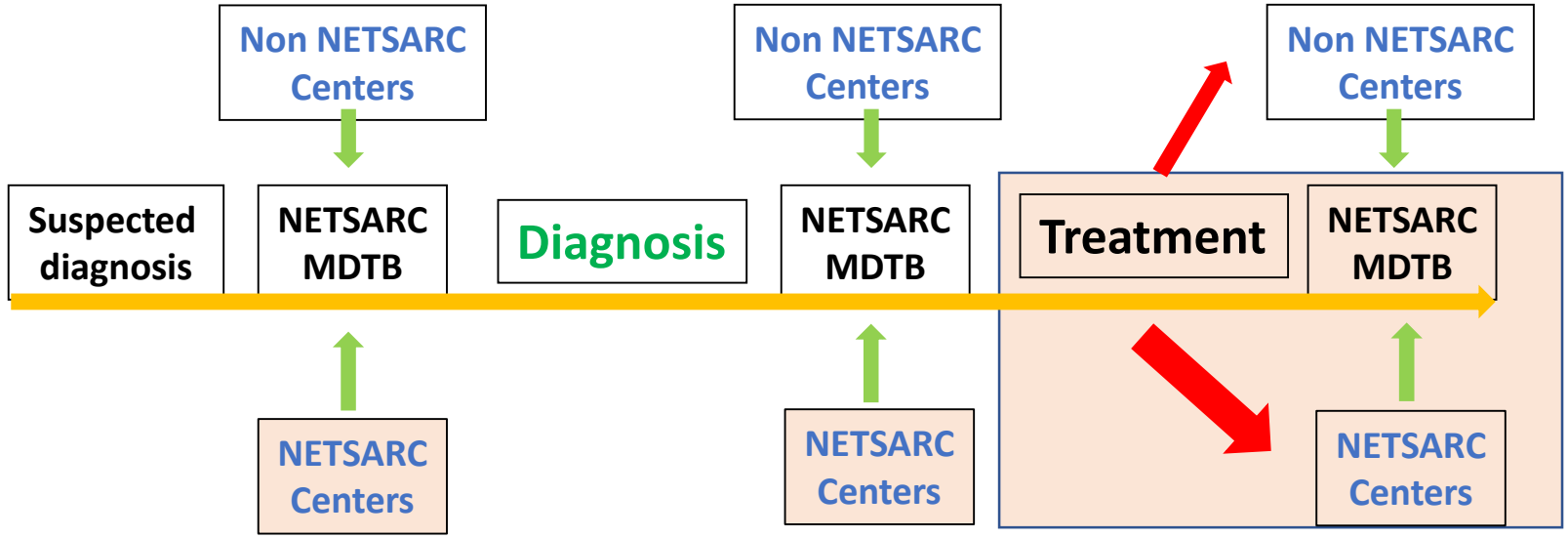
Annals of Oncology 30: 1143–1153, 2019
doi:10.1093/annonc/mdz124
Published online 9 April 2019

ORIGINAL ARTICLE

Surgery in reference centers improves survival of sarcoma patients: a nationwide study

J.-Y. Blay[1,2,3*], C. Honoré[4], E. Stoeckle[5], P. Meeus[1,2,3], M. Jafari[6,7], F. Gouin[1,2,3,8,9], P. Anract[10], G. Ferron[11], A. Rochwerger[12], M. Ropars[13,14], S. Carrere[15], F. Marchal[16], F. Sirveaux[16], A. Di Marco[17], L. R. Le Nail[18], J. Guiramand[19], G. Vaz[1,2,3], J.-C. Machiavello[20], O. Marco[21], S. Causeret[22], P. Gimbergues[23], F. Fiorenza[24], L. Chaigneau[25], F. Guillemin[26], J.-M. Guilloit[27], F. Dujardin[28], J.-P. Spano[29], J.-C. Ruzic[30], A. Michot[4], P. Soibinet[26], E. Bompas[8,9], C. Chevreau[11], F. Duffaud[12], M. Rios[13,14], C. Perrin[13,14], N. Firmin[15], F. Bertucci[19], C. Le Pechoux[4], F. Le Loarer[4], O. Collard[1,2,3], M. Karanian-Philippe[1,2,3], M. Brahmi[1,2,3], A. Dufresne[1,2,3], A. Dupré[1,2,3], F. Ducimetière[1,2,3], A. Giraud[10], D. Pérol[1,2,3], M. Toulmonde[5], I. Ray-Coquard[1,2,3], A. Italiano[5], A. Le Cesne[4], N. Penel[6,7] & S. Bonvalot[31], on behalf of the NETSARC/REPPS/RESOS and French Sarcoma Group–Groupe d'Etude des Tumeurs Osseuses (GSF-GETO) Networks†

- Does primary surgery the patient within a NetSARC center impacts survival?

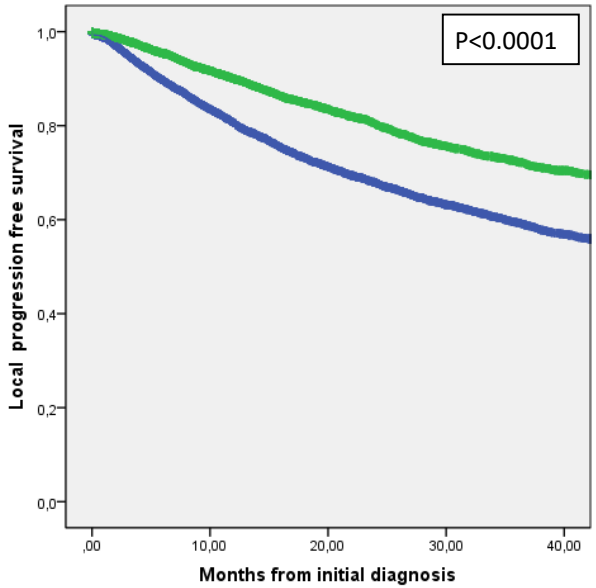


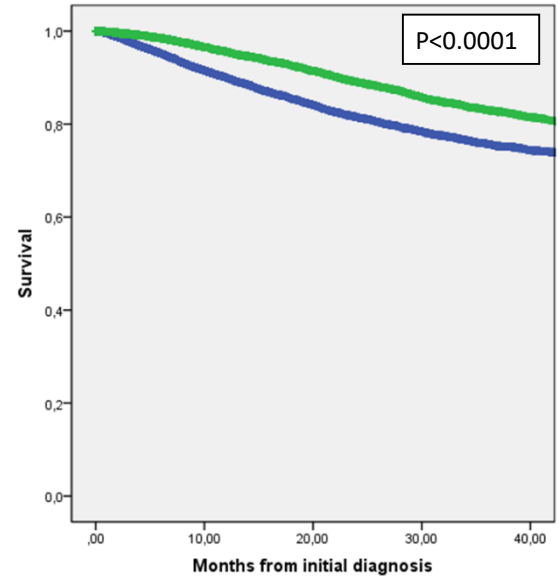
Quality of resection : initial & final

	Outside NETSARC or no data (N=19543)	Within NetSARC (N=9954)	p
Initial			
– **R0**	3114 (15.9%)	5280 (53.0%)	
– **R1**	3209 (16.%)	2388 (24.0%)	
– **R2**	1660 (8.5%)	417 (4.2%)	
– **UNK**	11560 (59.1%)	1869 (18.8%)	<0.000
Re-operation	2498 (21.4%)	616 (6.2%)	<0.000
Final			
– **R0**	4694 (24.0%)	5643 (56.7%)	
– **R1**	2493 (12.8%)	2170 (21.8%)	
– **R2**	982 (5.0%)	302 (3.0%)	
– **UNK**	11374 (58.2%)	1839 (18.5%)	<0.000

*: % of those with a date of surgery documented

LRFS & OS : incident patient population





Operated
- In NETSARC, N=9910 (33.9%)
- Outside NETSARC or
  no data, N=19307 (66.1%)

Multivariate analysis of prognostic factors for overall survival in the overall incident patient population of 29497 patients

	Beta	E.S.	Signif.	RR
Metastatic	1,207	,042	,000	3,342
NF1	,995	,143	,000	2,704
Size over 10	,506	,057	,000	1,658
Previous RT	,470	,082	,000	1,599
Deep	,290	,043	,000	1,336
Grade 3	,204	,046	,000	1,226
Gender	,161	,038	,000	1,175
Previous cancer	,128	,057	,023	1,137
Age at diagnosis	,016	,001	,000	1,016
Grade 2	-,328	,055	,000	,720
Surgery Netsarc	-,451	,042	,000	,637
Grade 1	-1,174	,103	,000	,309
GIST	-1,704	,117	,000	,182
Intermediate malignancy	-2,078	,138	,000	,125

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Annals of
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ORIGINAL ARTICLE – SARCOMA

Survival Benefit of the Surgical Management of Retroperitoneal Sarcoma in a Reference Center: A Nationwide Study of the French Sarcoma Group from the NetSarc Database

S. Bonvalot, MD, PhD[1], E. Gaignard, MD[1], E. Stoeckle, MD[2], P. Meeus, MD[3], G. Decanter, MD[4], S. Carrere, MD[5], C. Honore, MD, PhD[6], J. B. Delhorme, MD[7], M. Fau, MD[8], D. Tzanis, MD[1], S. Causeret, MD[9], P. Gimbergues, MD[10], J. M. Guillois, MD[11], B. Meunier, MD[12], A. Le Cesne, MD[13], F. Ducimetiere, PhD[14], M. Toulmonde, MD, PhD[15], and J. Y. Blay, MD, PhD[16]

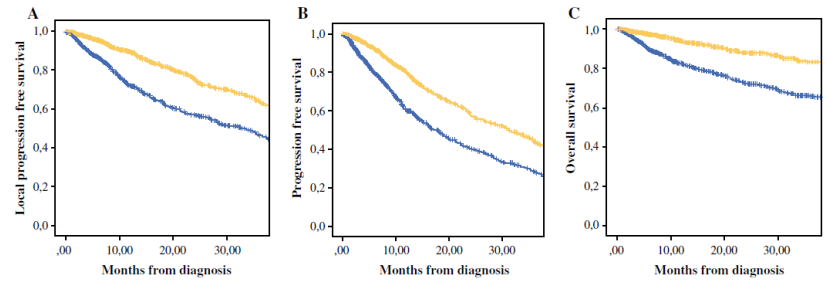

FIG. 1 Local progression-free survival (**a**), progression-free survival (**b**), and overall survival (**c**) of the retroperitoneal sarcoma patients in the NetSarc database. In blue: patients operated on outside of the NetSarc network. In yellow: patients operated on at a NetSarc center. Log-rank $p < 0.0001$ for LRFS, PFS, and OS

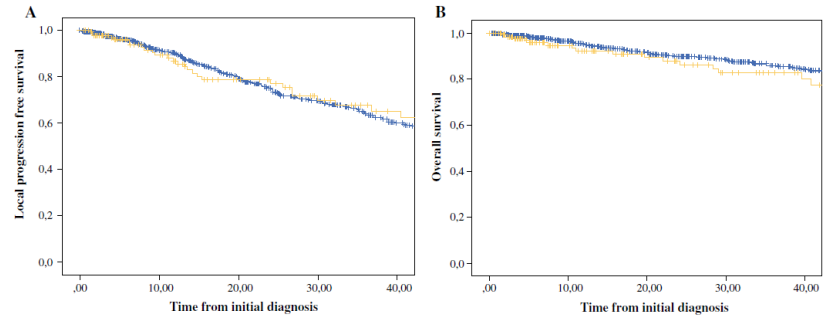

FIG. 2 Local progression-free survival (**a**) and overall survival (**b**) of patients operated on at NetSarc centers. In blue: patients operated on at the 13 NetSarc centers with the largest accrual of RPS patients. In yellow: patients operated on at the 13 NetSarc centers with the smallest accrual of RPS patients. Log-rank $p > 0.05$

Limits of this analysis

3 groups		**% meta**	**% RFS@ 2yrs** (non meta)
– Pts operated in NETSARC	N=9954	675 (6.5%)	66%
– Pts operated outside NETSARC	N=11671	895 (7.7%)	59%
– Pts without date of surgery	N=7872	1924 (24.4%)	60%

- Surgery in NETSARC center also an independent prognostic factor for LRFS, RFS and OS in non metastatic patients.
- Surgery in NETSARC center also an independent prognostic factor for LRFS, and RFS in group 1 vs Group 2
- « Surgery in a NETSARC center is associated with a better outcome »

Topics

- DSCRT
- General guidelines
- Local treatments, surgery, HIPEC, WART…
- Systemic treatments

EJSO (2005) 31, 917-923

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the Journal of Cancer Surgery
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Randomized trial of cytoreduction followed by intraperitoneal chemotherapy versus cytoreduction alone in patients with peritoneal sarcomatosis

S. Bonvalot[a,*], A. Cavalcanti[a], C. Le Péchoux[b], P. Terrier[c], D. Vanel[d], J.Y. Blay[e], A. Le Cesne[f], D. Elias[a]

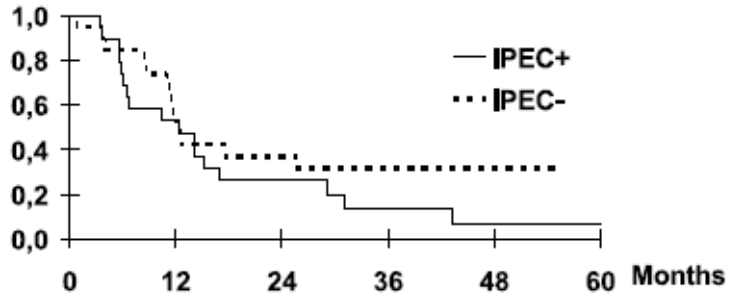

Figure 1 Local recurrence free survival (IPEC intraperitoneal chemotherapy).

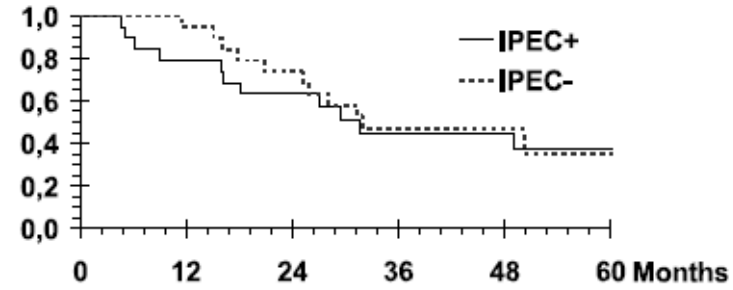

Figure 3 Overall survival (IPEC intraperitoneal chemotherapy).

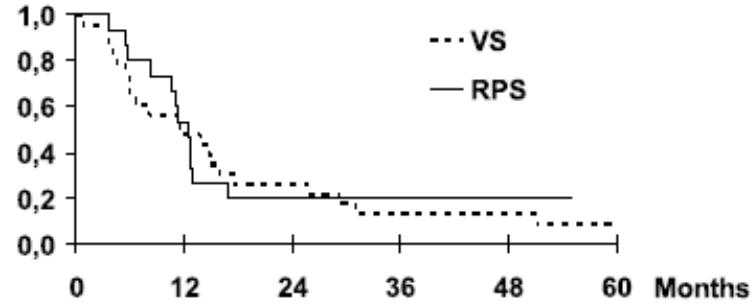

Figure 2 Metastases free survival (VS, visceral sarcoma; RPS, retroperitoneal sarcoma).

Surgical Oncology 29 (2019) 107–112

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Table 1
Characteristics at diagnosis.

Variable		n = 100 (100%)
Sex		
	Male	81 (81%)
	Female	19 (19%)
Mean age, years [SD]		27 [12]
Median age, years [range]		25 [3–59]
WHO performance status		
	0	65 (65%)
	1	17 (17%)
	2	4 (4%)
	N/A	14 (14%)
Median PCI [range]		13 [2–30]
Extra-peritoneal metastases		25(25%)
	Liver metastases	20(20%)
	Lung metastases	3(3%)
	Bone metastases	3(3%)
	Other	3(3%)
More than one EPM site		15 (15%)
MD Anderson stage		
	I	27(27%)
	II	24(24%)
	III	13(13%)
	IV	7(7%)
	N/A	30(30%)
Symptoms		89 (89%)
	Pain	57 (57%)
	Occlusion	2 (2%)
	Bloating	11 (11%)
	Other	19 (19%)
Time of referral to expert center		
	Before any biopsy	18 (18%)
	After initial diagnosis	26 (26%)
	Before surgery	12 (12%)
	After treatment	16 (16%)
	After recurrence	4 (4%)
	Other/unknown	24 (24%)

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Table 2
Treatments.

Variable		n = 100 (100%)
Induction chemotherapy		
	Yes	80 (80%)
	No	20 (20%)
Surgery		
	Yes	71 (71%)
	No	29 (29%)
Completeness of cytoreductive surgery		
	CC0	46 (46%)
	CC1	4 (4%)
	CC2/3	21 (21%)
Intraperitoneal chemotherapy		17 (17%)
	HIPEC	15 (15%)
	EPIC	2 (2%)
Intraperitoneal chemotherapy drug		
	Cisplatin	10 (10%)
	Oxaliplatin	6 (6%)
	MMC	6 (6%)
Postoperative WAP-RT		26 (26%)
Postoperative chemotherapy		54 (54%)
	Pre- and postoperative	36 (36%)

Abbreviations: HIPEC, hyperthemic intraperitoneal; chemotherapy; EPIC, early postoperative intraperitoneal chemotherapy; MMC, mitomycin; WAP-RT, whole abdomino-pelvic radiotherapy.

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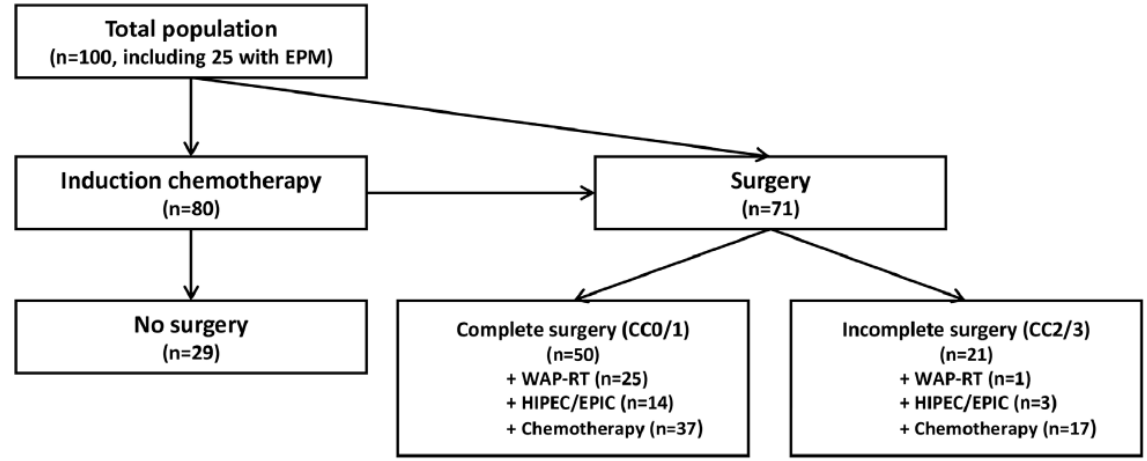

Abbreviation : EPM, extra-peritoneal metastases; WAP-RT, whole abdomino-pelvic radiotherapy; HIPEC, hyperthemic intraperitoneal chemotherapy; EPIC, early postoperative intraperitoneal chemotherapy

Fig. 1. Selection process and treatment flowchart.

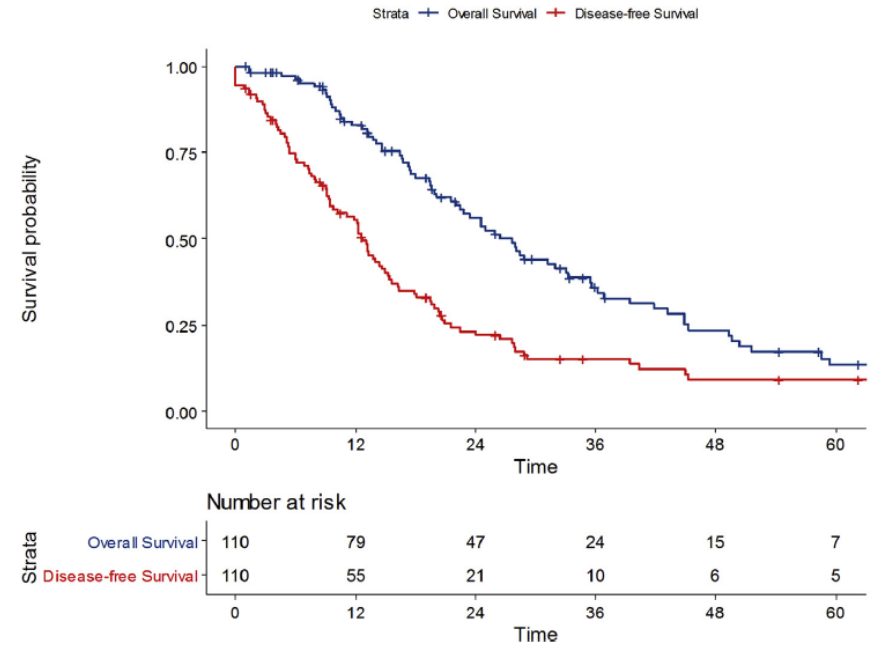

Fig. 2. Overall and disease free survival.

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Table 3
Predictive factors for cure (univariate analysis).

	Not cure, n = 95 (100%)	Cured, n = 5 (100%)	p
Sex			0.005
Male	80 (84%)	1 (20%)	
Female	15 (16%)	4 (80%)	
Median age, years [range]	25 [3–59]	28 [16–48]	0.62
WHO performance status			0.19
0	61 (64%)	4 (80%)	
1	17 (18%)	0 (0%)	
2	4 (4%)	0 (0%)	
N/A	13 (14%)	1 (20%)	
Median PCI [range]	13 [2–30]	6 [2–9]	0.005
Extra-peritoneal metastases	25 (26%)	0 (0%)	0.19
Liver metastases	20 (21%)	0 (0%)	0.35
Lung metastases	13 (14%)	0 (0%)	0.59
Bone metastases	3 (3%)	0 (0%)	1
Other	3 (3%)	0 (0%)	1
More than one EPM site	15 (15%)	0 (0%)	0.60
MD Anderson stage			0.003
I	22 (23%)	5 (100%)	
II	24 (25%)	0 (0%)	
III	13 (14%)	0 (0%)	
IV	7 (7%)	0 (0%)	
N/A	30 (31%)	0 (0%)	
Symptoms at diagnosis	85 (89%)	4 (80%)	1
Time of referral to expert center			0.51
Before any biopsy	18 (19%)	0 (0%)	
After initial diagnosis	25 (26%)	1 (20%)	
Before surgery	10 (11%)	1 (20%)	
After treatment	16 (17%)	0 (0%)	
After recurrence	3 (4%)	1 (20%)	
Other/unknown	23 (23%)	2 (40%)	
Completeness of cytoreductive surgery			0.005
CC0/1	45 (48%)	5 (100%)	
CC2/3	50 (52%)	0 (0%)	
HIPEC	15 (16%)	0 (0%)	0.65
Preoperative chemotherapy	47 (48%)	4 (80%)	1
Postoperative chemotherapy	53 (56%)	1 (20%)	1
Pre- and postoperative	35 (37%)	3 (38%)	0.47
Postoperative WAP-RT	22 (23%)	4 (80%)	0.003

Abbreviation: PCI, peritoneal cancer index; WAP-RT, EPM, extra-peritoneal metastases; WAP-RT, whole abdomino-pelvic radiotherapy.

Published OnlineFirst June 5, 2018; DOI: 10.1158/1078-0432.CCR-18-0202

Translational Cancer Mechanisms and Therapy

Clinical Cancer Research

Multimodality Treatment of Desmoplastic Small Round Cell Tumor: Chemotherapy and Complete Cytoreductive Surgery Improve Patient Survival

Vivek Subbiah[1], Salah-Eddine Lamhamedi-Cherradi[1], Branko Cuglievan[2], Brian A. Menegaz[1], Pamela Camacho[2], Winston Huh[2], Vandhana Ramamoorthy[1], Pete M. Anderson[3], Raphael E. Pollock[4], Dina C. Lev[4], Wei Qiao[5], Mary Frances McAleer[6], Robert S. Benjamin[1], Shreyaskumar Patel[1], Cynthia E. Herzog[2], Najat C. Daw[2], Barry W. Feig[7], Alexander J. Lazar[8], Andrea Hayes-Jordan[7], and Joseph A. Ludwig[1]

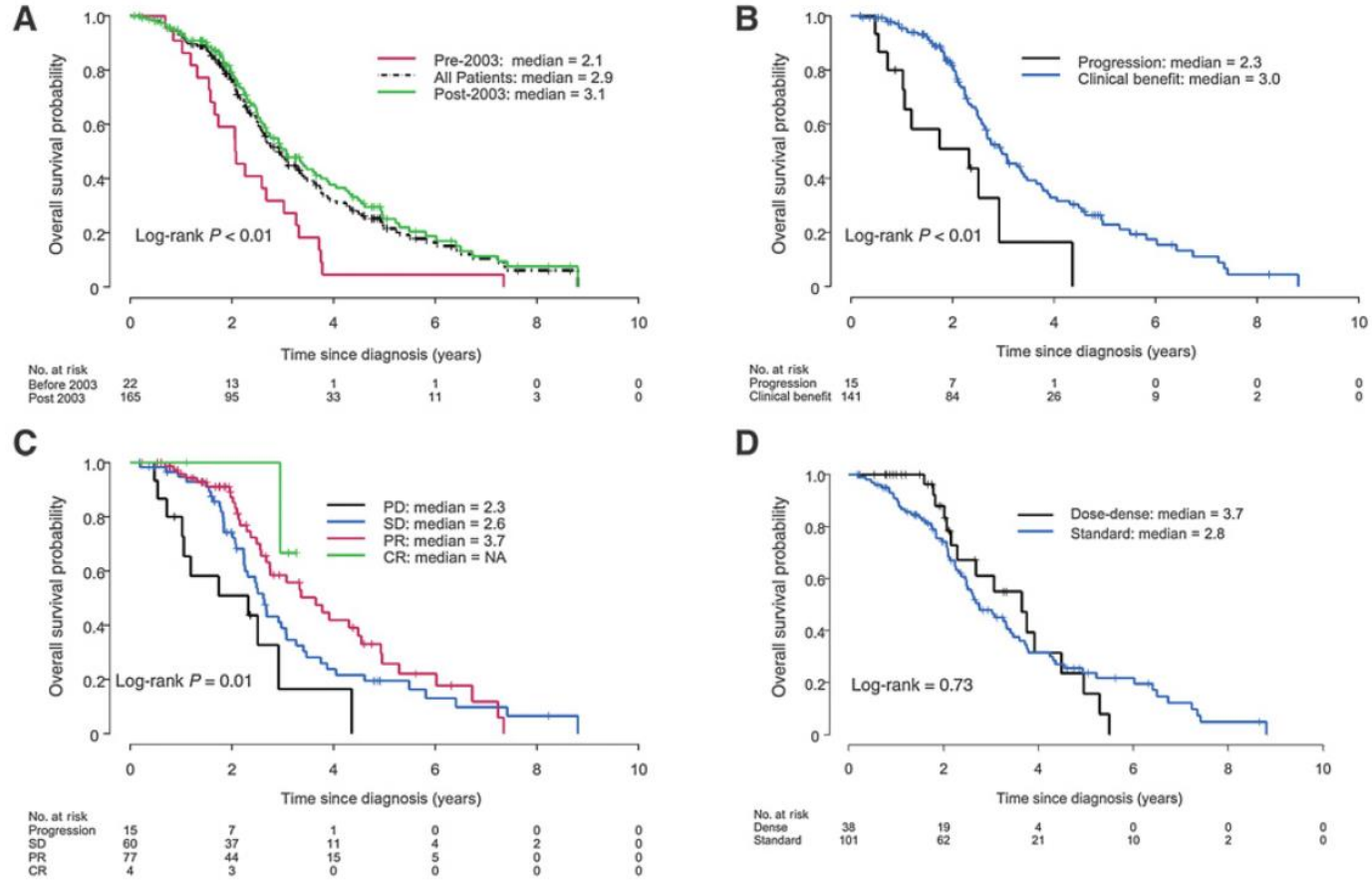

Figure 1.

Chemotherapy impact upon OS in patients with DSRCT. **A,** OS for all patients according to treatment era (before 2003, after 2003, and both). **B,** OS in patients who achieved clinical benefit or had progressive disease to their initial chemotherapy regimen. **C,** Stratification of OS by RECIST response to the first chemotherapy regimen each patient received. (**D**) Survival effect resulting from standard 3-week chemotherapy treatment vs. dose-dense treatment, when the traditional neoadjuvant VAC/IE regimen was provided. CR, complete response; PD, partial disease; PR, partial response; SD, stable disease.

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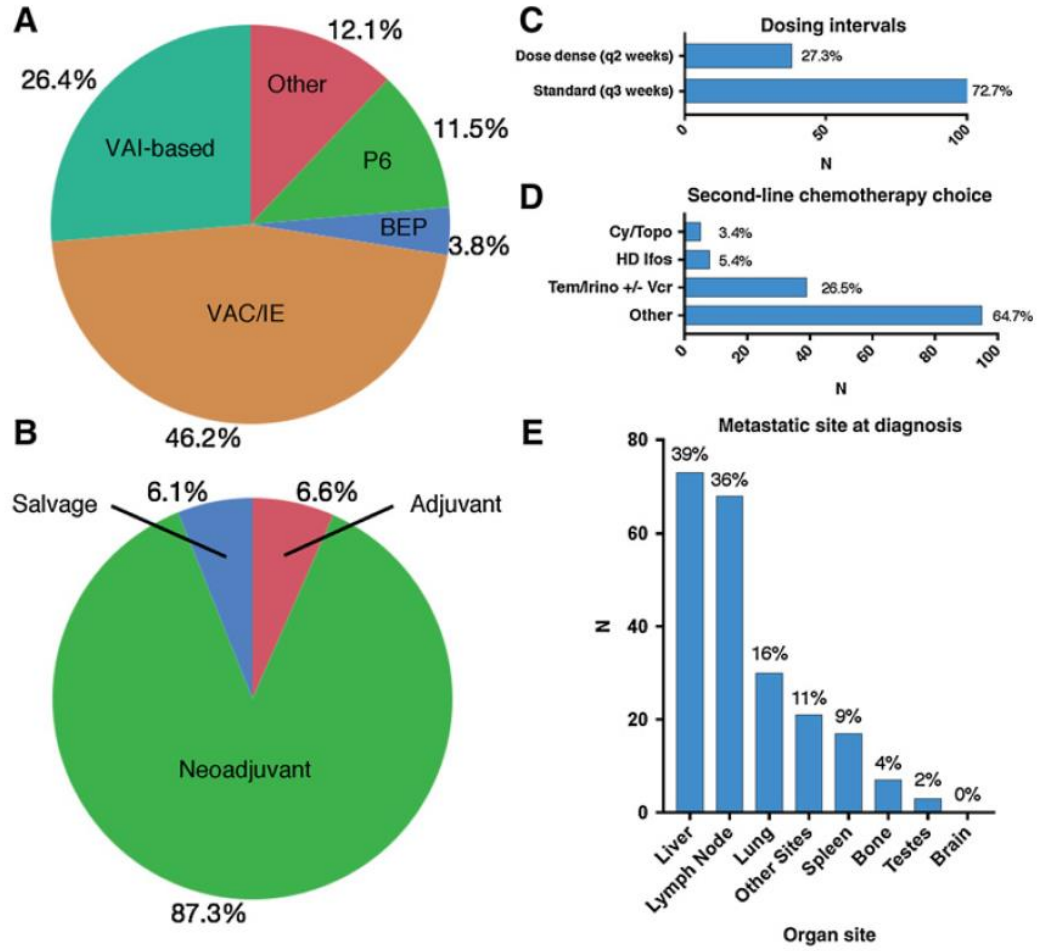

Figure 2.

Chemotherapy patterns in patients with DSRCT. **A,** The initial chemotherapy regimens used to treat patients with DSRCT. **B,** Chemotherapy administration in relation to surgery: included neoadjuvant (87.3%), adjuvant (6.6%), or salvage (6.1%) treatments. **C,** Dosing interval in patients with DSRCT. 72.7% of the patients received standard therapy every 3 weeks, and 27.3% received dense therapy every 2 weeks. **D,** Second-line chemotherapy treatment regimens used in patients with DSRCT. **E,** Organ sites involved among the 114 patients who presented with metastases at the time of diagnosis. P6, seven cycles of IE/VDC (Ifosfamide, Etoposide, Vincristine, Adriamycin, and Cytoxan); BEP, bleomycin, etoposide, and cisplatin; VAI, Vincristine, actinomycin, and ifosfamide; Cy, cyclophosphamide; Topo, topotecan; HD ifos, high-dose ifosfamide; Tem, temozolomide; Irino, irinotecan; Vcr, vincristine; *N*, number of metastatic sites.

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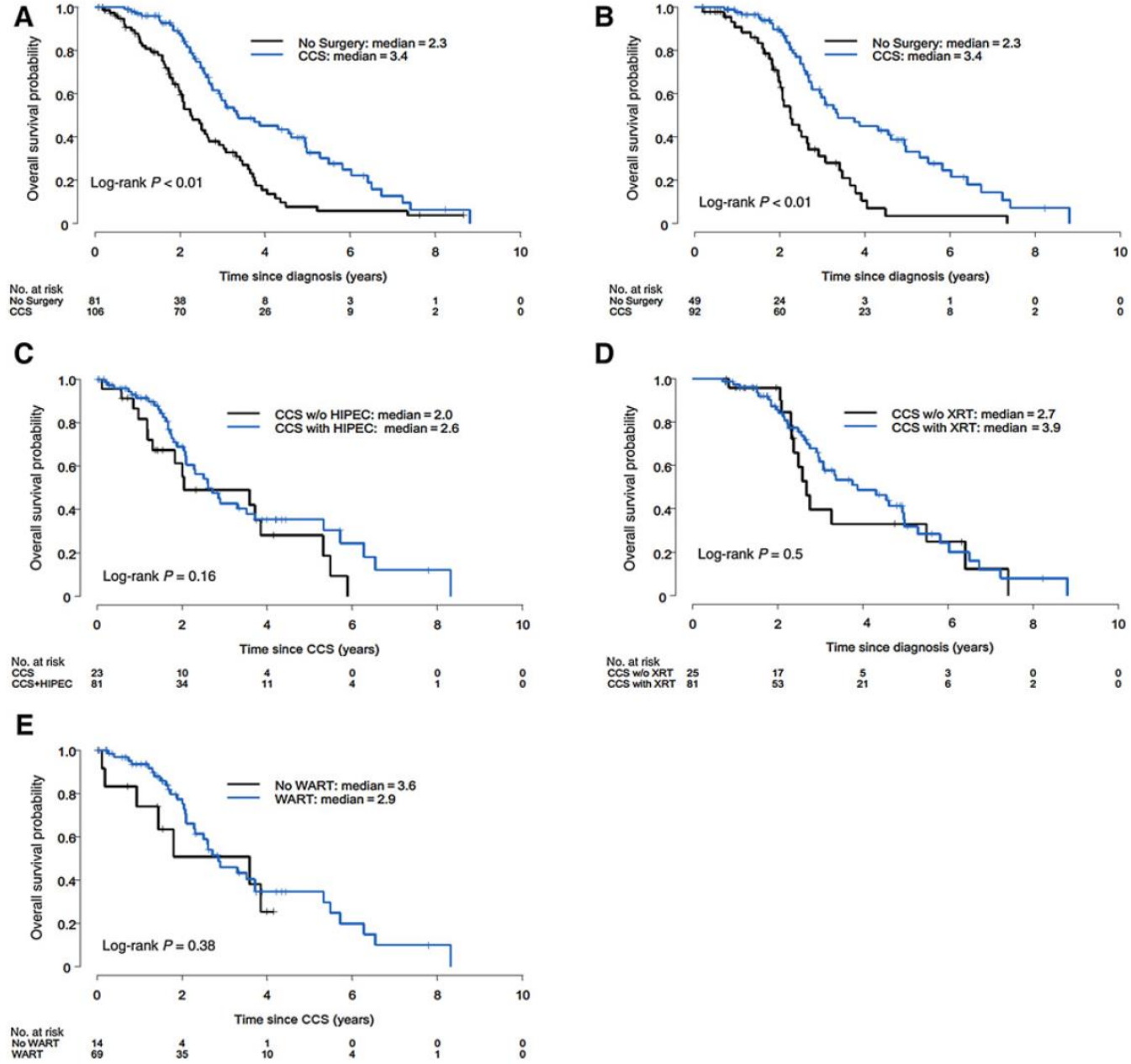

Figure 3.

Impact upon OS by surgery, HIPEC, radiation, and WART. **A,** OS in patients who underwent CCS. **B,** OS benefit of CCS in patients who achieved clinical benefit from their first neoadjuvant chemotherapy regimen. **C,** OS in patients that underwent CCS ± HIPEC. **D,** OS in patients who received radiation at some time during their clinical care. **E,** The OS effect of WART, which was almost always provided 6 to 8 weeks following CCS.

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Annals of
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ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Desmoplastic Small Round Cell Tumor Treated with Cytoreductive Surgery and Hyperthermic Intraperitoneal Chemotherapy: Results of a Phase 2 Trial

Andrea A. Hayes-Jordan, MD[1], Brian A. Coakley, MD[1], Holly L. Green, PA-C[2], LianChun Xiao, MS[3], Keith F. Fournier, MD[1], Cynthia E. Herzog, MD[4], Joseph A. Ludwig, MD[5], Mary F. McAleer, MD, PhD[6], Peter M. Anderson, MD, PhD[7], and Winston W. Huh, MD[4]

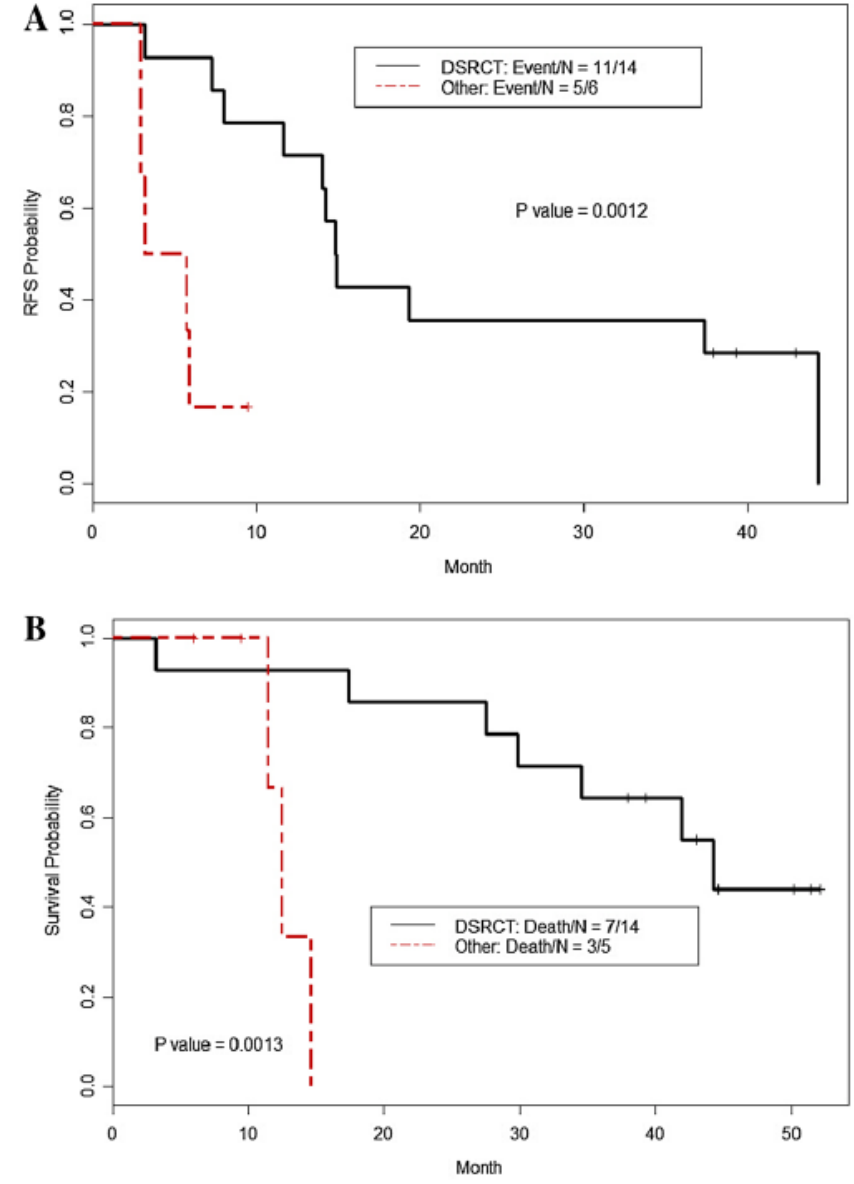

FIG. 2 a Recurrence-free and b overall survival in DSRCT versus other sarcoma tumors after CRS-HIPEC with cisplatin

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Translational Cancer Mechanisms and Therapy

Clinical Cancer Research

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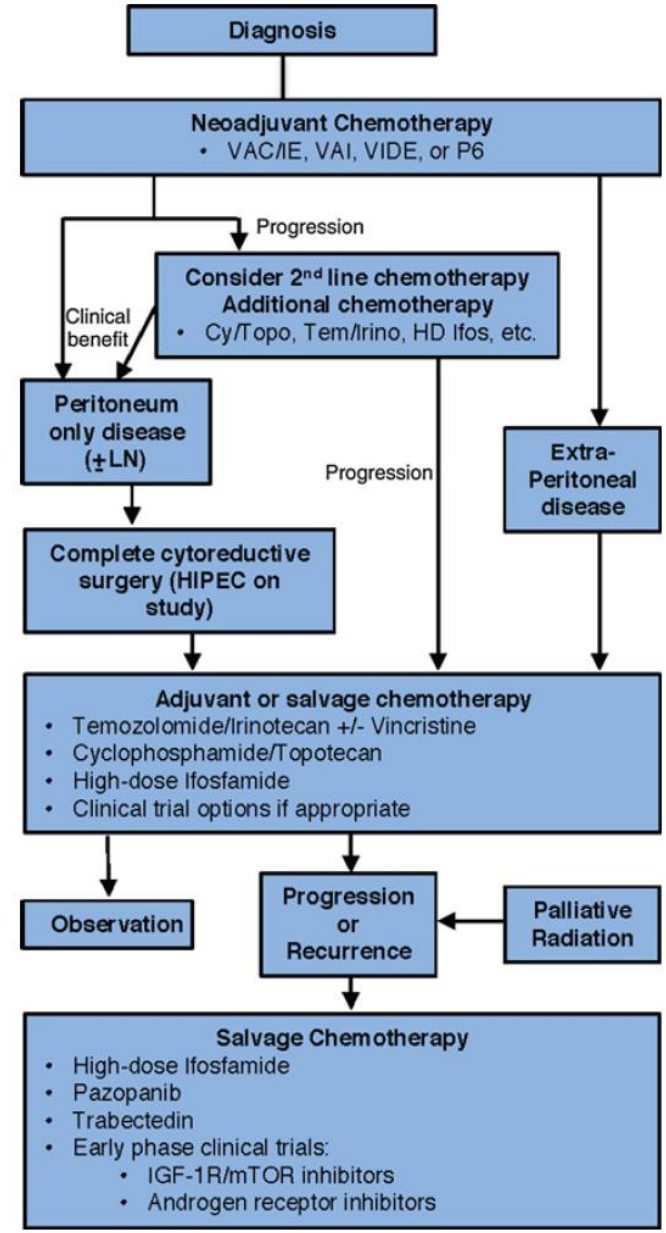

Figure 4.

Revised DSRCT treatment algorithm adapted by MDACC. The diagnosis of DSRCT is confirmed using IHC and/or cytology, and the staging workup is

Topics

- DSCRT
- General guidelines
- Local treatments, surgery, HIPEC, WART…
- Systemic treatments

Cytotoxics, targeted treatments and immunotherapy

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ELSEVIER

Trabectedin in patients with advanced soft tissue sarcoma: A retrospective national analysis of the French Sarcoma Group

Axel Le Cesne [a,*], Isabelle Ray-Coquard [b], Florence Duffaud [c], Christine Chevreau [d], Nicolas Penel [e], Binh Bui Nguyen [f], Sophie Piperno-Neumann [g], Corinne Delcambre [h], Maria Rios [i], Loic Chaigneau [j], Christine Le Maignan [k], Cecile Guillemet [l], François Bertucci [m], Emmanuelle Bompas [n], Claude Linassier [o], Thimotée Olivier [p], Jean-Emmanuel Kurtz [q], Caroline Even [a], Philippe Cousin [b], Jean Yves Blay [b], for the French Sarcoma Group

Table 2
Progression-free survival and overall survival per type of sarcoma histology.

Histology	*N*	PFS (months)			OS (months)	
		Median	95% CI	PFS at 3 months (%)	Median	95% CI
Solitary fibrous tumour	13	7.633	1.593–13.673	69	14.333	.839–27.827
Chondrosarcoma	13	6.267	.000–15.935	57	21.400	9.641–33.159
Liposarcoma	161	6.067	4.488–7.645	64	15.000	11.033–18.967
Leiomyosarcoma	321	5.500	4.528–6.472	69	15.133	13.350–16.917
Fibrosarcoma	10	5.400	2.508–8.292	70	13.733	7.230–20.237
Epithelioid sarcoma	10	4.633	2.498–6.768	70	12.033	8.328–15.738
Synovial sarcoma	101	3.933	2.090–5.776	53	9.867	6.738–12.995
DSCRT	5	3.400	.000–9.126	60	14.000	.000–34.827
Myxofibrosarcoma	20	2.833	1.471–4.196	47	8.100	4.790–11.410
Stromal sarcomas	14	2.800	1.700–3.900	43	12.767	8.234–17.299
Rhabdomyosarcoma	15	2.567	.757–4.376	47	5.433	4.606–6.261
Sarcoma NOS	82	2.367	1.977–2.756	42	6.367	4.614–8.120
MPNST	19	2.367	1.040–3.694	42	7.767	3.317–12.217
Miscellaneous	89	2.300	.968–3.632	46	7.967	6.664–9.270
Osteosarcoma	3	1.967	.000–5.061	33	6.400	.000–13.175
Angiosarcoma	9	.933	.836–1.031	22	6.567	.519–12.615

CI; confidence intervals; DSCRT, Desmoplastic Small Round Cell Tumour; MPNST, Malignant Peripheral Nerve Sheath Tumours; OS, overall survival; PFS, progression-free survival.

ANTICANCER RESEARCH *30*: 547-552 (2010)

Phase II Clinical Trial of Imatinib Mesylate in Therapy of KIT and/or PDGFRα-expressing Ewing Sarcoma Family of Tumors and Desmoplastic Small Round Cell Tumors

JOSEPH CHAO[1], G. THOMAS BUDD[2], PEIGUO CHU[3], PAUL FRANKEL[4], DOLORES GARCIA[4], MARIBEL JUNQUEIRA[1], SOFIA LOERA[3], GEORGE SOMLO[1], JUDITH SATO[5] and WARREN A. CHOW[1]

OPEN ACCESS Freely available online

PLOS ONE

Morphoproteomic Profiling of the Mammalian Target of Rapamycin (mTOR) Signaling Pathway in Desmoplastic Small Round Cell Tumor (EWS/WT1), Ewing's Sarcoma (EWS/FLI1) and Wilms' Tumor(WT1)

Vivek Subbiah[1*], Robert E. Brown[2], Yunyun Jiang[1], Jamie Buryanek[2], Andrea Hayes-Jordan[3], Razelle Kurzrock[1,4], Pete M. Anderson[5]

VOLUME 30 · NUMBER 15 · MAY 20 2012

JOURNAL OF CLINICAL ONCOLOGY

Phase II Study of Ganitumab, a Fully Human Anti–Type-1 Insulin-Like Growth Factor Receptor Antibody, in Patients With Metastatic Ewing Family Tumors or Desmoplastic Small Round Cell Tumors

William D. Tap, George Demetri, Phillip Barnette, Jayesh Desai, Petr Kavan, Richard Tozer, Pasquale W. Benedetto, Gregory Friberg, Hongjie Deng, Ian McCaffery, Ian Leitch, Sunita Badola, Sung Chang, Min Zhu, and Anthony Tolcher

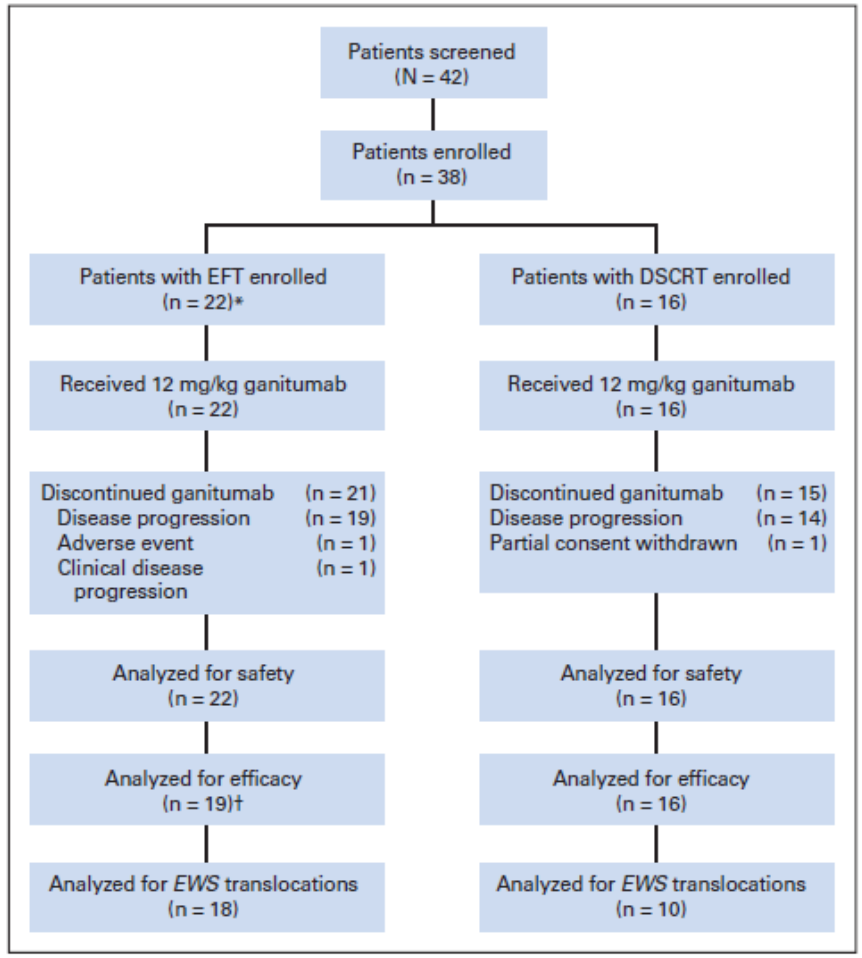

Table 1. Baseline Demographics and Disease Characteristics

Characteristic	Primary Cohort: Ewing Family Tumor* (n = 19) No.	%	Primary Cohort: Desmoplastic Small Round Cell Tumor (n = 16) No.	%	Exploratory Cohort Ewing Family Tumor* (n = 3) No.	%	Total (N = 38) No.	%
Sex								
Female	6	32	1	6	1	33	8	21
Male	13	68	15	94	2	67	30	79
Race/ethnicity								
White	15	79	10	63	3	100	28	74
Black or African American	1	5	1	6	0	0	2	5
Hispanic or Latino	2	11	4	25	0	0	6	16
Asian	1	5	1	6	0	0	2	5
Age, years								
Median	29		33		30		30	
Range	20-77		19-63		25-30		19-77	
ECOG score								
0	7	37	9	56	0	0	16	42
1	9	47	7	44	3	100	19	50
2	3	16	0	0	0	0	3	8
Disease stage								
Stage II	1	5	0	0	1	33	2	5
Stage III	1	5	3	19	0	0	4	11
Stage IV	14	74	11	69	2	67	27	71
Unknown	3	16	2	13	0	0	5	13
No. of lesions†								
1	3		3		0		6	
2	8		2		1		11	
3 to 5	5		4		1		10	
> 5	3		7		0		10	
Metastatic sites‡								
Lung	11		2		1		14	
Liver	3		8		0		11	
Mediastinum	4		6		1		11	
Node	6		6		0		12	
Mesentery	0		1		0		1	
Adrenal	2		2		1		5	
Other§	6		11		1		18	
Lines of prior cancer therapy								
1	6	32	2	13	0	0	8	21
2	5	26	6	38	1	33	12	32
≥ 3	8	42	8	50	2	67	18	47
Lines of prior radiotherapy								
0	6	32	11	69	1	33	18	47
1	8	42	4	25	1	33	13	34
2	2	11	1	6	1	33	4	11
≥ 3	3	16	0	0	0	0	3	8

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Table 2. Best Tumor Response

Response	Ewing Family Tumor		Desmoplastic Small Round Cell Tumor		Total	
	No.	%	No.	%	No.	%
Patients with measurable disease at baseline	19	100	16	100	35	100
Response assessment						
PR	1	5	1	6	2	6
SD	7	37	10	63	17	49
SD ≥ 24 weeks	1	5	3	18	4	12
Clinical benefit (PR + SD ≥ 24 weeks)	2	11	4	25	6	17
Progressive disease	10	53	4	25	14	40
Not evaluable	1	5	1	6	2	6

Abbreviations: PR, partial response; SD, stable disease.

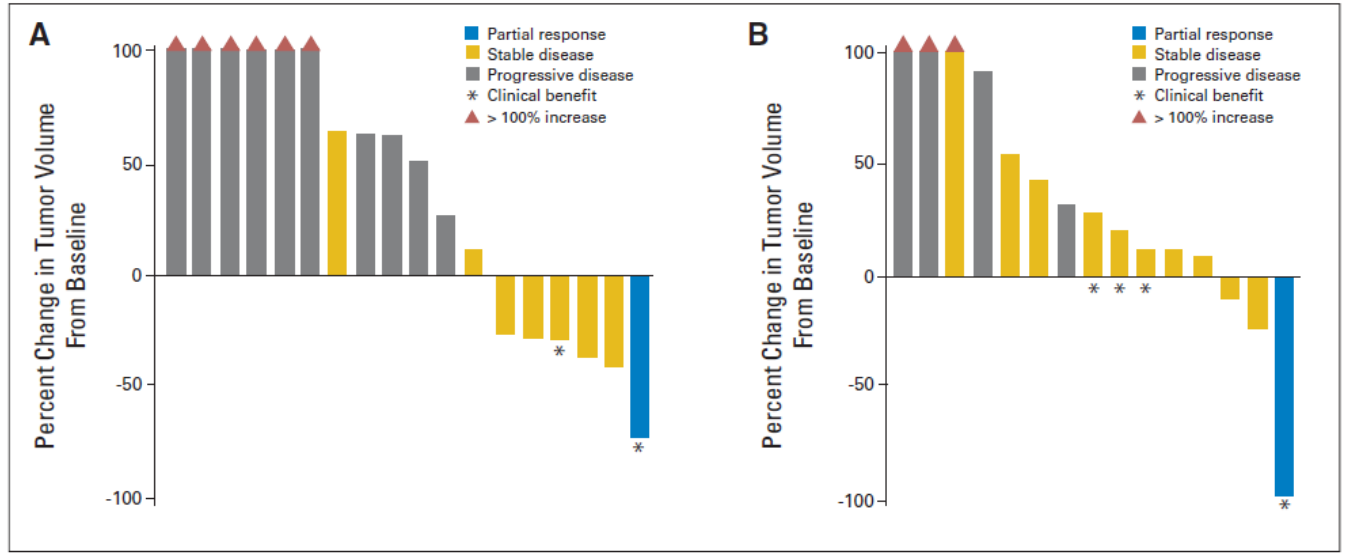

Fig 2. Best percentage change in sum of tumor volumes from baseline in Ewing family tumor (A) and desmoplastic small round cell tumor (B). Best response measured by RECIST. (*) Patients with clinical benefit (partial response or stable disease ≥ 24 weeks).

The off-label use of targeted therapies in sarcomas: the OUTC'S program

Lauriane Eberst[1,2], Claire Cropet[1], Axel Le Cesne[3], Patricia Pautier[3], Nicolas Penel[4], Antoine Adenis[4], Christine Chevreau[5], Jacques-Olivier Bay[6,7], Olivier Collard[8], Didier Cupissol[9], Florence Duffaud[10], Jean-Claude Gentet[10], Sophie Piperno-Neumann[11], Perrine Marec-Berard[1], Emmanuelle Bompas[12], Antoine Thyss[13], Loic Chaigneau[14], Philippe Cassier[1], François Bertucci[15], Jean-Yves Blay[1,2] and Isabelle Ray-Coquard[1,2*]

Table 2 Targeted therapy by histotypes

Targeted therapy	N	%	Histotype 1	n (%)	Histotype 2	n (%)	Histotype 3	n (%)	Histotype 4	n (%)	Histotype 5	n (%)
Sorafenib (1)	125	45	GIST	31 (25)	LMS	22 (18)	AS	14 (11)	Uterine LMS	8 (6)	Liposarcoma	8 (6)
Suntinib (2)	67	24	LMS	9 (13)	Ewing	8 (12)	SS	8 (12)	Unclassified S	8 (12)	Uterine LMS	4 (6)
Imatinib	23	8	Chordoma	8 (35)	AF	4 (17)	DFSP	4 (17)	Epithelioid S	2 (9)	—	—
Sirolimus-cyclophosphamide	18	6	OsteoS	8 (44)	ChondroS	5 (27)	AS/chordoma/lipoS/Ewing/SFT	1 each (6)	—	—	—	—
Everolimus (3)	10	4	GIST	3 (30)	LMS	3(30)	KS/MPNST/SS	1 each (10)	Other	1(10)	—	—
Bevacizumab (4)	9	3	Other	5 (56)	MFST	2 (22)	AS	1 (11)	Epithelioid S	1 (11)	—	—
Sirolimus alone	5	2	OsteoS	2 (40)	PEComa	1 (20)	other	1 (20)	—	—	—	—

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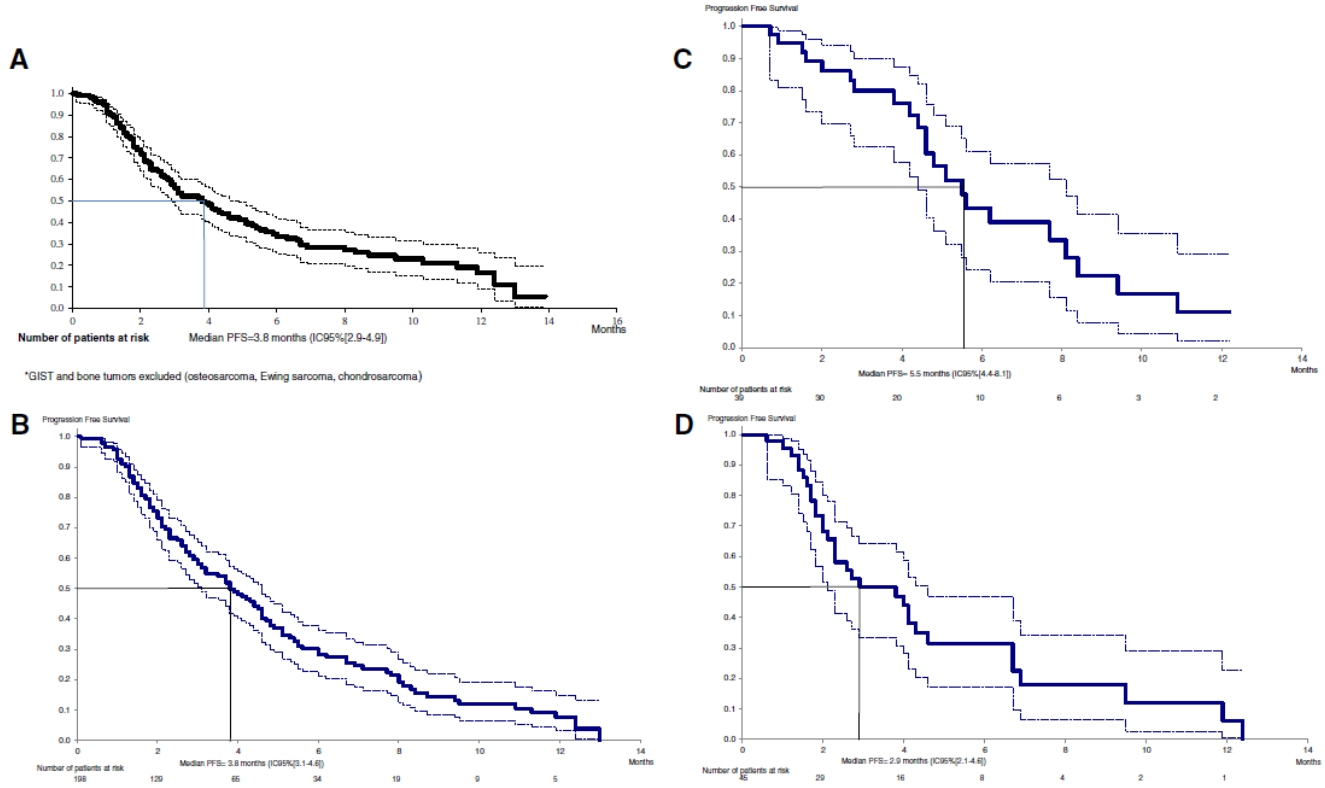

Figure 1 Progression-free survival data. A. Progression-free survival for soft-tissue sarcoma (except GIST). **B**: Progression-free survival of the patients in third line and after (number of previous chemotherapy >2). **C**: Progression-free survival of the GIST group. **D**: Progression-free survival of the leiomyosarcoma group.

Bétrian et al. Clin Sarcoma Res (2017) 7:10
DOI 10.1186/s13569-017-0076-4

RESEARCH Open Access

Antiangiogenic effects in patients with progressive desmoplastic small round cell tumor: data from the French national registry dedicated to the use of off-labeled targeted therapy in sarcoma (OUTC's)

Sarah Bétrian[1*], Christophe Bergeron[2], Jean-Yves Blay[3], Emmanuelle Bompas[4], Philippe A. Cassier[3], Laure Chevallier[5], Jérome Fayette[3], Magali Girodet[5], Cécile Guillemet[6], Axel Le Cesne[7], Perrine Marec-Berard[2], Isabelle Ray-Coquard[3] and Christine Chevreau[1]

Table 4 Literature data of chemotherapy agents and TT for advanced TT

Chemotherapy agent	Patients number	Best response	PFS	References
Vinorelbine-Cyclophosphamide	2	2 PR	4 and 15 months	Ferrari et al. [18]
Irinotecan	8	4 SD, 4 PD	Unknown	Bisogno et al. [19]
Trabectedine	2	2 SD	4 months	Frezza et al. [21]
TT agent				
Imatinib	7	1 PR, 4 PD, 2 NA	1 month	Chao et al. [22]
Sunitinib	8	2 PR, 3 SD, 3 PD	2.6 months	Italiano et al. [23]
Pazopanib[c]	9	2 PR, 5 SD, 2 PD, CBR[a] 78%	9.2 months	Frezza et al. [24]
Temsirolimus	1	SD	10 months	Thijs et al. [33]
Ganitumab	16	1 PR, 10 SD, 4 PD, CBR[b] 25%	19 months	Tap et al. [30]

PR partial response, *SD* stable disease, *PD* progressive disease, *NA* non assessable, *CBR* clinical benefit rate (PR + SD >12 weeks[a], 24 weeks[b])

[c] Retrospective analysis from data comprising three DSRCT patients treated within the EORTC phase II study 62043, three in the EORTC phase III study 62072 (PALETTE), along with three patients treated in the UK on the subsequent pazopanib named patient program

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Clinical Sarcoma Research

RESEARCH Open Access

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Sarah Bétrian[1*], Christophe Bergeron[2], Jean-Yves Blay[3], Emmanuelle Bompas[4], Philippe A. Cassier[3], Laure Chevallier[5], Jérome Fayette[3], Magali Girodet[5], Cécile Guillemet[6], Axel Le Cesne[7], Perrine Marec-Berard[2], Isabelle Ray-Coquard[3] and Christine Chevreau[1]

Table 2 Treatment response and duration

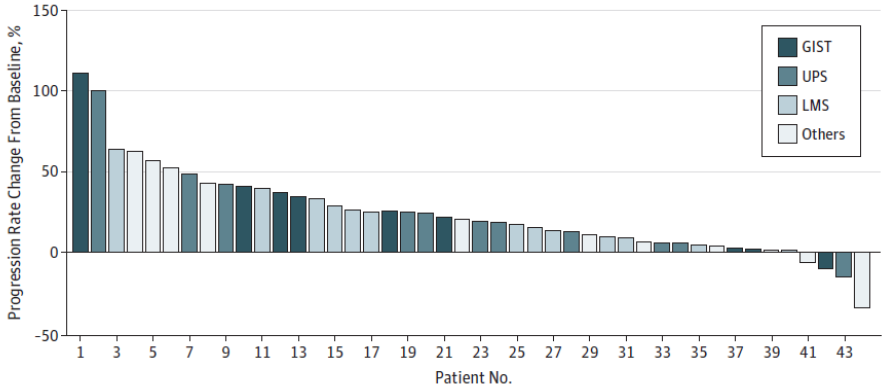
Patient	First treatment	Surgery resection quality	Radiotherapy	PFS 1	Relapses treatments/*PFS*	Lines of treatment before TT	TT type	PFS with TT (months)
1	MAID[a]-ASCT[b]	R2	_	12 months	Gemcitabine-cisplatine/2,5 months	2	Sunitinib	2
2	LV5Fu-Ciplatine	_	_	8 months	HIPEC[c]-FOLFIRI[d]/17 months FOLFIRI/3 months Holoxan-etoposide/3 months Adriamycine-cyclophosphamide/*2 months*	*5*	Sorafenib	3.5
3	MAI[e]	R1	Yes	39 months	Gemcitabine-docetaxel/4 months Adriamycine-holoxan/3 months Cisplatine-irinotecan/6 months Trabectedine/9 months	5	Sorafenib	4
4	Adriamycine-etposide-Cisplatine-Cyclophosphamide + VAI[f]	R2	_	44 months	Carboplatine-etoposide/13 months	2	Sunitinib	5.5
5	Cyclophosphamide-etoposide-carboplatine	R1	Yes	47 months	Etoposide-carboplatine-busulfan-thiotepa/12 months Temozolamide/5 months TEMIRI[g]/*4 months*	4	Bevacizumab	2
6	MAI	_	_	6 months	VAC[h]/6 months	2	Ridaforolimus *PFS* 23 months Sunitinib	4 OS 38 months
7[j]	Bevacizumab-IVADo[i]	R1	Yes	7 months	Navelbine-cyclophosphamide/10 months	2	Dalotuzumab *PFS* 2 months Sunitinib	3
8	Adriamycine-ifosfamide-etoposide	_	_	6 months	Cyclophosphamide/3 months Trabectedine/4 months	3	Sunitinib	2 OS 19 months
9	MAI	_	_	7 months	VAC/3 months	2	Sunitinib	2

JAMA Oncology | Brief Report

Use of PD-1 Targeting, Macrophage Infiltration, and IDO Pathway Activation in Sarcomas
A Phase 2 Clinical Trial

Maud Toulmonde, MD; Nicolas Penel, MD, PhD; Julien Adam, MD, PhD; Christine Chevreau, MD; Jean-Yves Blay, MD, PhD; Axel Le Cesne, MD; Emmanuelle Bompas, MD; Sophie Piperno-Neumann, MD; Sophie Cousin, MD; Thomas Grellety, MD; Thomas Ryckewaert, MD; Alban Bessede, PhD; François Ghiringhelli, MD, PhD; Marina Pulido, MSc; Antoine Italiano, MD, PhD

Figure 2. Change in Progression Rate for 44 Patients



Article

B cells are associated with survival and immunotherapy response in sarcoma

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Florent Petitprez[1,2,3,4], Aurélien de Reyniès[4,24], Emily Z. Keung[5,24], Tom Wei-Wu Chen[6,7,8,9], Cheng-Ming Sun[1,2,3], Julien Calderaro[1,10,11], Yung-Ming Jeng[9,12], Li-Ping Hsiao[7], Laetitia Lacroix[1,2,3], Antoine Bougoüin[1,2,3], Marco Moreira[1,2,3], Guillaume Lacroix[1,2,3], Ivo Natario[1,2,3], Julien Adam[13], Carlo Lucchesi[14,15], Yec'han Laizet[14,15], Maud Toulmonde[14,16], Melissa A. Burgess[17], Vanessa Bolejack[18], Denise Reinke[19], Khalid M. Wani[20], Wei-Lien Wang[20], Alexander J. Lazar[20,21], Christina L. Roland[5], Jennifer A. Wargo[5,21], Antoine Italiano[14,16,22], Catherine Sautès-Fridman[1,2,3], Hussein A. Tawbi[23]* & Wolf H. Fridman[1,2,3]*

Article

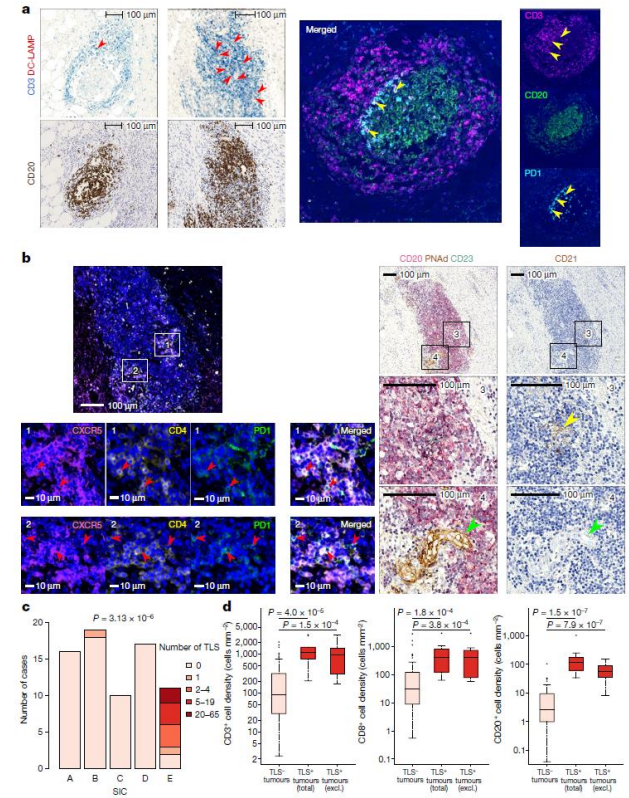


Fig. 3 | TLSs are a distinguishing feature of the immune-high class of STS. right) positive cells with reticular morphology characteristic of follicular

Axitinib plus pembrolizumab in patients with advanced sarcomas including alveolar soft-part sarcoma: a single-centre, single-arm, phase 2 trial

Breelyn A Wilky, Matteo M Trucco, Ty K Subhawong, Vaia Florou, Wungki Park, Deukwoo Kwon, Eric D Wieder, Despina Kolonias, Andrew E Rosenberg, Darcy A Kerr, Efrosyni Sfakianaki, Mark Foley, Jaime R Merchan, Krishna V Komanduri, Jonathan C Trent

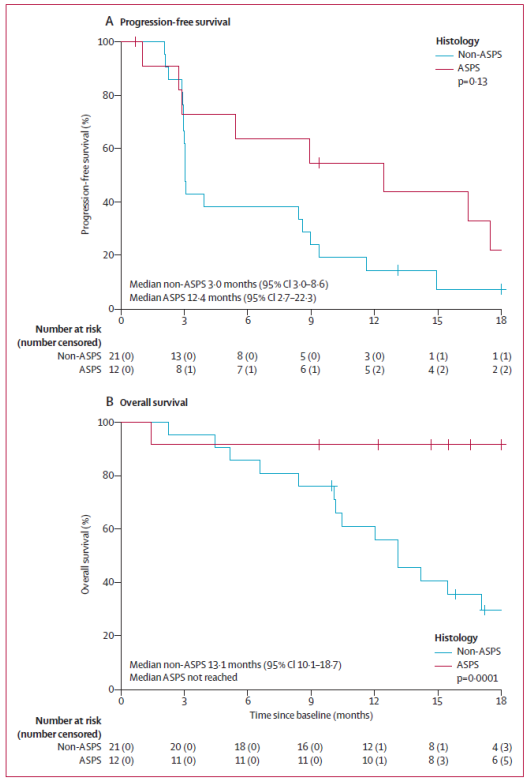


Figure 5: **Post-hoc analysis of progression-free survival (A) and overall survival (B) by ASPS status**
Kaplan-Meier estimates by intention-to-treat analysis (n=33). Patients who remained event-free were censored at the time of last imaging or withdrawal of consent. We compared curves from ASPS and non-ASPS populations using the log-rank test. Crosses indicate censored patients. ASPS=alveolar soft part sarcoma. NE=non-estimable.

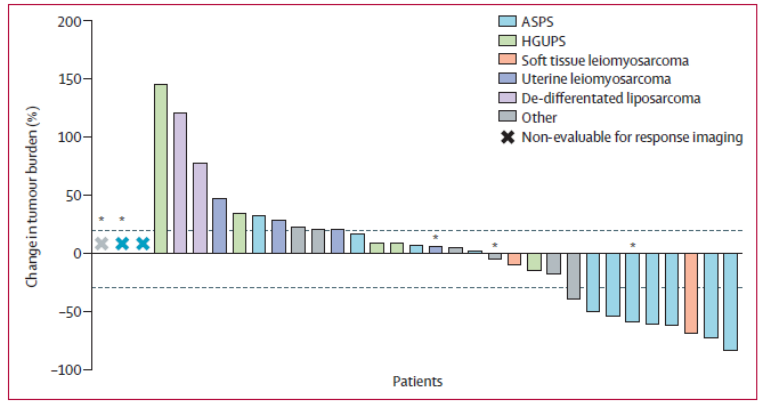


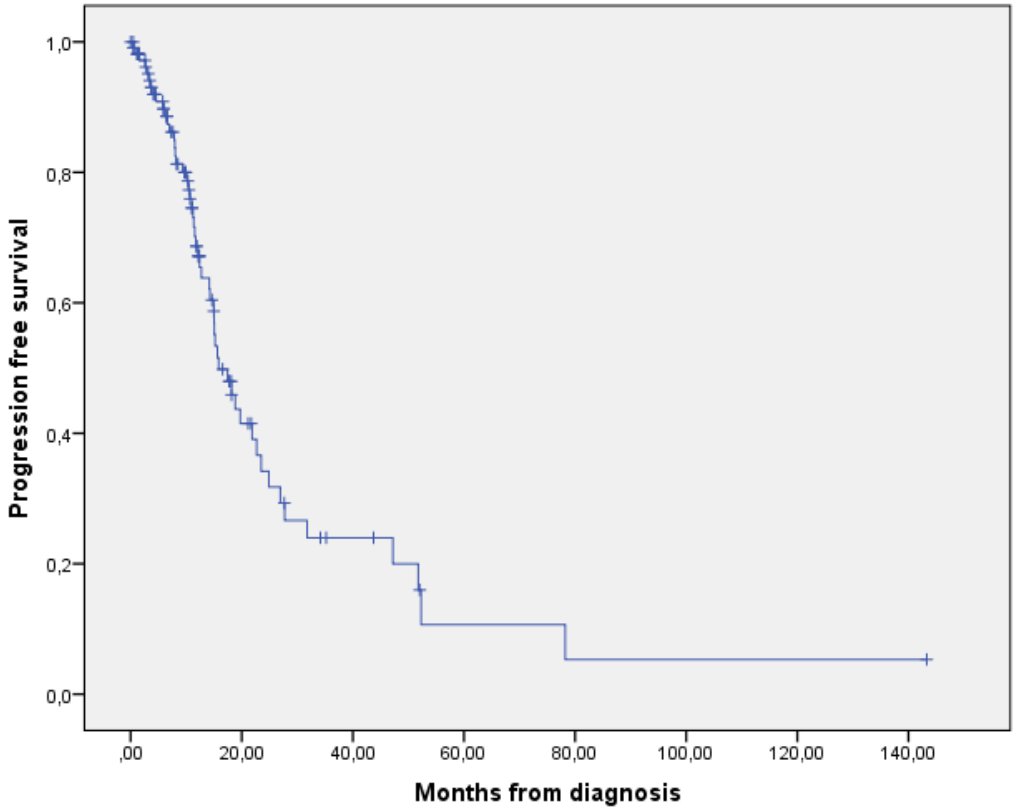
Figure 3: **Change from baseline in tumour burden**

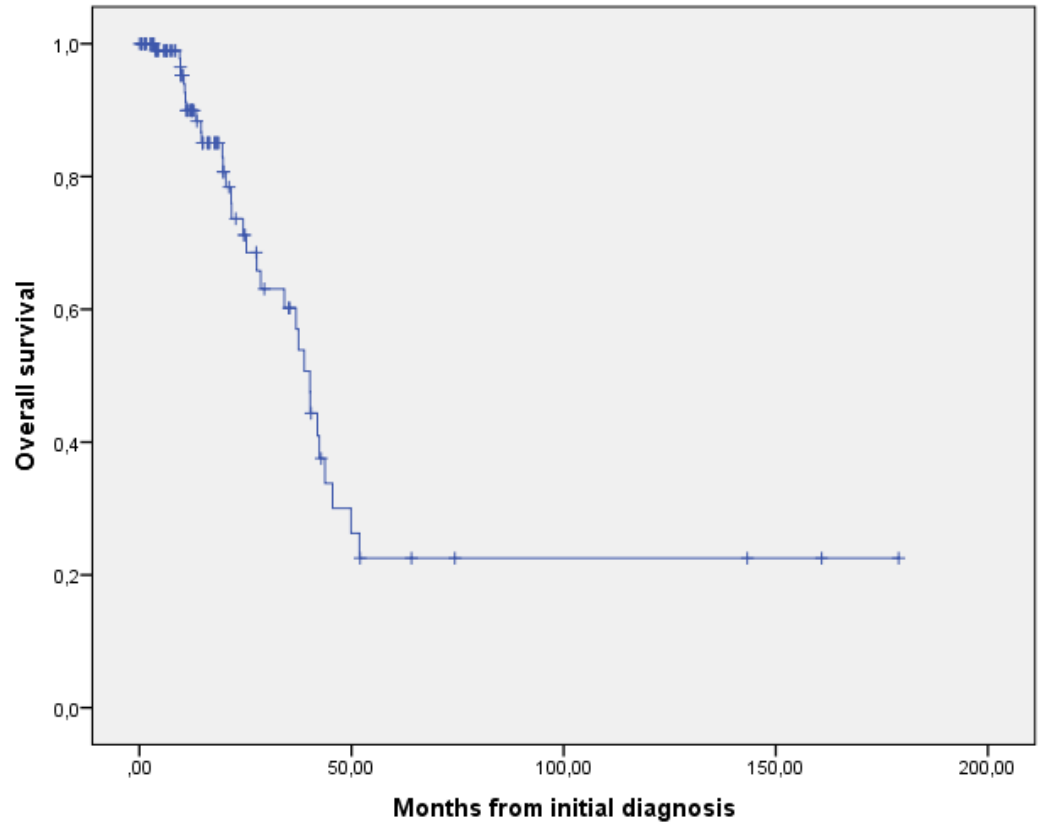
ACSE Pembrolizumab
(C. Massard, A. Marabelle)

Sarcoma arm

- ASPS
- Chordomas
- Rhabdoid
- DSCRT

DSCRT : NETSARC series, N=113





Conclusions

- Very rare
- Quality of primary management and multidisciplinarity
- Neoadjuvant CT, surgery, adjuvant CT
- Cytoreductive surgery
- WA-RT?
- HIPEC?

- New agents needed