AIEOP
ASSOCIAZIONE ITALIANA EMATOLOGIA ONCOLOGIA PEDIATRICA

Presidente
A. Mastronuzzi

Segreteria Scientifica
A. Colombini, T. Perillo, A. Zibaldo

Giornate AIEOP

RIMINI
Hotel Savoia

13-14 aprile 2026

Sarcomi Parti Molli

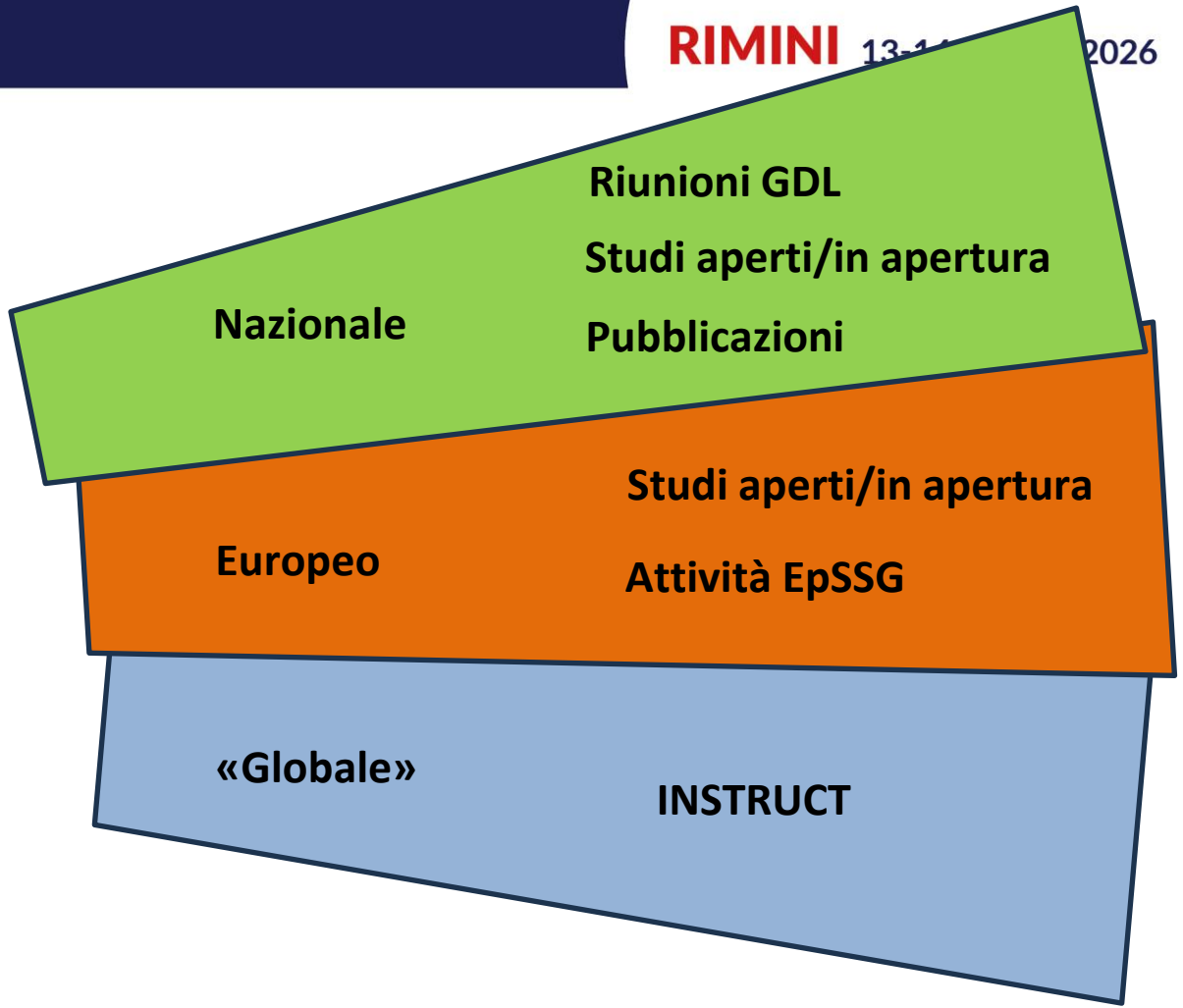
Gianni Bisogno

Dipartimento di Salute della Donna e del Bambino
Padova

Disclosures

Company name	Research support	Employee	Consultant	Stockholder	Speakers bureau	Advisory board	Other

Associazione Italiana Ematologia Oncologia Pediatrica
STSC Soft Tissue Sarcoma Committee



Incontri del GDL

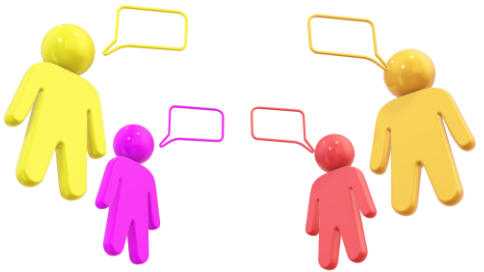
Membri
- Gianni Bisogno
- Simona Affinita
- Stefano Chiaravalli
- Francesco De Leonardis
- Carla Manzitti
- Giuseppe Maria Milano
- Katia Perruccio

Consulenti
- Rita Alaggio
- Michela Casanova
- Andrea Ferrari
- Elena Poli

Membri ~~Allargati~~ Interessati
- Patrizia Bertolini
- Paolo Bonvini
- Federica Cennamo
- Daniela Di Carlo
- Valentina Di Ruscio
- Veronica Folsi
- Virginia Livellara
- Federico Mercolini
- Evelina Miele
- Ida Russo
- Luisa Santoro
- Pietro Soloni
- Stefania Cardellicchio
- ……

Altri
- Laboratori
- Study coordinator
- Data Manager

Quando
- 14/4/2025
- 08/05/2025
- 28/07/2025
- 10/10/2025
- 12/12/2025
- 24/02/2026



Progetto SAPERE (PNRR)

Promotore	OPBG – AIEOP – PI: prof.ssa Alaggio
Centri Italiani	4 (OPBG, PD, BA, PA)
Protocollo	Versione 1.0 del 29/06/2024
Tipo di Studio	Osservazionale prospettivo
Stato dello studio	Approvazione CE OPBG 26/07/2024 – Convenzione col Ministero firmata – Accordi con UO in firma – Firmato Accordo Studio Specifico Padova 02/2026
Durata studio	08/2024 – 08/2026 → proroga di 6 mesi
Criteri di inclusione	• 0 – 21 anni • Diagnosi istologica di sarcoma parti molli
Pazienti da reclutare	150 pz
Aggiornamento	Piattaforma eCRF in sviluppo
Criticità	Lo studio non può essere aperto in altri centri italiani

Studio AIEOP SPM

Promotore	AIEOP (AOUP) – PI: dott.ssa Affinita
Centri Italiani	4/36 centri attivati: OPBG, Bari Policlinico, Padova, Palermo Arnas
Protocollo	Versione 1.0 del 02/11/2023
Tipo di studio	Osservazionale prospettico
Stato dello studio	Approvazione CET 12/2024 – Convenzione firmata – Arruolamento in corso (PD)
Durata studio	2024 – 2034
Pazienti eligibili	• 0 – 21 anni • Tutti i SPM
Pazienti da reclutare	150/anno in Italia
Pazienti reclutati	9 pz arruolati
Aggiornamento	• In attesa implementazione Dbase
Prossimi passi	• Attivare i centri italiani • Informare AIEOP se è cambiato il PI locale

Studio Fertilità

Promotore	AIEOP (AOUP) – PI: dott.ssa Affinita
Centri Italiani	8/14 – centri attivati: Bari (4 pz), Firenze, Genova, INT Milano (4), Parma (1), Padova (5), Perugia(3), Verona
Protocollo	Versione 1.0 del 02/11/2023
Tipo di studio	Osservazionale prospettico
Stato dello studio	Approvazione CET 12/2024 – Convenzione firmata – Arruolamento in corso
Durata studio	2024 – 2027
Pazienti eligibili	• Pazienti puberi maschi • RMS
Pazienti da reclutare	54
Pazienti reclutati	17
Aggiornamento	• Approvato da CE, in attesa contratto: Bologna, Catania Napoli, Napoli. • Sottomesso al CE: Bergamo, Torino • In attesa: OPBG
Prossimi passi	• Continua arruolamento

RASopatie e RMS pediatrico con NF1

Rhabdomyosarcoma arising in patients with *RAS*opathies: a distinct subgroup with peculiar features? S. Vallese[1], S. Scuderi[2], I. Giovannoni[1], C. Tancredi[1], M.C. Affinita[3], G. Bisogno[3], G.M. Milano[4], A.P. Dei Tos[5], R. Alaggio[1], L. Santoro[5]	
Background	• Multiple genetic conditions predispose to RMS, including RASopathies (risk 20 times greater than in the general population) • Clinically defined group of medical genetic syndromes caused by germline mutations in genes of the Ras/MAPK pathway: - crucial role in cell cycle, cellular growth, differentiation and senescence; - dysregulation lead deleterious effects on both embryonic and later stages of development • These disorders include: **Neurofibromatosis type 1 (NF1), Noonan syndrome (NS), Noonan syndrome with multiple lentigines (NSML), Costello syndrome (CS), cardio-facio-cutaneous syndrome (CFCS), capillary malformation-AVM syndrome (CM-AVM), Legius syndrome** • About 40 cases of RMS in in RASopathies described in leterature (20 NF1; 3 Noonan Syndrome; 15 Costello Syndrome; 2 CFCS) • Age range: 0-13 yrs; slight male predominance; GU localization (22 cases), followed by head/ neck, biliary tract, died of disease (7/32, 21%) 3 died of other causes • Histology: mainly ERMS; 2 ARMS in Costello syndrome

Valore predittivo della [18F]FDG-PET nel Rabdomiosarcoma

Promotore	OPBG – Ida Russo/Federico Mercolini
Tipo studio	Osservazionale, retrospettivo, multicentrico
Periodo	2017 – 2024
Background clinico	La risposta precoce alla chemioterapia di induzione è un indicatore prognostico riconosciuto in diversi tumori pediatrici (ES, OS, NB). Nel RMS, la valutazione radiologica convenzionale (TC, RM) ha dimostrato basso valore predittivo per la sopravvivenza libera da eventi (EFS). Studi IRS-IV e COG D9803 hanno evidenziato questo limite
Obiettivi dello studio	**Primario:** Valutare il ruolo di **MTV e TLG** prima e dopo la chemioterapia di induzione come predittori di EFS (Event-Free Survival); LFS (Local Failure Survival); OS (Overall Survival) **Secondari:** Correlare **SUL/SUV, MTV e TLG** con la risposta secondo criteri **RECIST**
Popolazione	Identificati 70 potenziali pazienti

Possibili nuovi studi/iniziative

- Cancer Predisposition syndrome (GM Milano)
- RMS Specific tumour primary site
- Outcome e Trattamento pazienti in recidiva
- NRSTS cosa succede dopo la recidiva
- Malattia minima (sangue, midollo, altri fluidi) – primo draft in preparazione

- Network dei laboratori (Padova – Milano – Roma)

Pubblicazioni GDL SPM 2025-26

Diagnostic Delay and Survival in Pediatric Rhabdomyosarcoma: Is Time a Critical Factor?

Affinita MC, Chiaravalli S, Milano GM, Russo I, Perruccio K, Tagarelli A, Bertolini P, Manzitti C, Mercolini F, Tamburini A, De Leonardis F, D'Angelo P, Cardellicchio S, Di Cataldo A, Di Pinto D, Mura RM, Coppadoro B, Ferrari A, **Bisogno G.**

J Pediatr Hematol Oncol. 2026 Jan 1;48(1):e15-e20. doi: 10.1097/MPH.0000000000003150. Epub 2025

Early radiologic tumour volume response in non-metastatic rhabdomyosarcoma is not predictive for survival.

de Vries ISA, Morosi C, **Bisogno G**, Minard-Colin V, Coppadoro B, Zanetti I, Ferrari A, Orbach D, Moalla S, Ben-Arush M, Devalck C, van Ewijk R, McHugh K, Jenney M, Chisholm J, Guillen G, Mandeville H, Merks JHM, Van Rijn RR.

Pediatr Radiol. 2025 Sep;55(10):2160-2170. doi: 10.1007/s00247-025-06359-3. Epub 2025 Aug 14.

Assessment of Lung Nodules in Children With Pediatric Sarcoma Undergoing [18F]-FDG-PET/MR for Staging.

Fichera G, Cecchin D, Stramare R, **Bisogno G**, Causin F, Zucchetta P, Giraudo C.

Pediatr Blood Cancer. 2025 May;72(5):e31622. doi: 10.1002/pbc.31622. Epub 2025 Feb 24.

Systemic therapy in metastatic pediatric rhabdomyosarcoma: a history of challenges and the search for promising approaches.

Di Carlo D, Affinita MC, Poli E, **Bisogno G.**

Expert Opin Pharmacother. 2025 Apr;26(6):755-763. doi: 10.1080/14656566.2025.2484319. Epub 2025 Mar 31.

Accettato per pubblicazione

Characteristics and outcome of patients with refractory orbital rhabdomyosarcoma

Gianni Bisogno [1,2], Maria Carmen Affinita [2], Giuseppe Maria Milano [3], Ilaria Zanetti [2], Elena Poli [2], Francesco De Leonardis [4], Katia Perruccio [5], Rita Alaggio [6], Stefano Chiaravalli [7], Virginia Livellara [8,9], Giovanni Scarzello [10], Andrea Ferrari [7,11]

Nazionale

Protocolli chiusi

Protocolli aperti/in apertura

Europeo

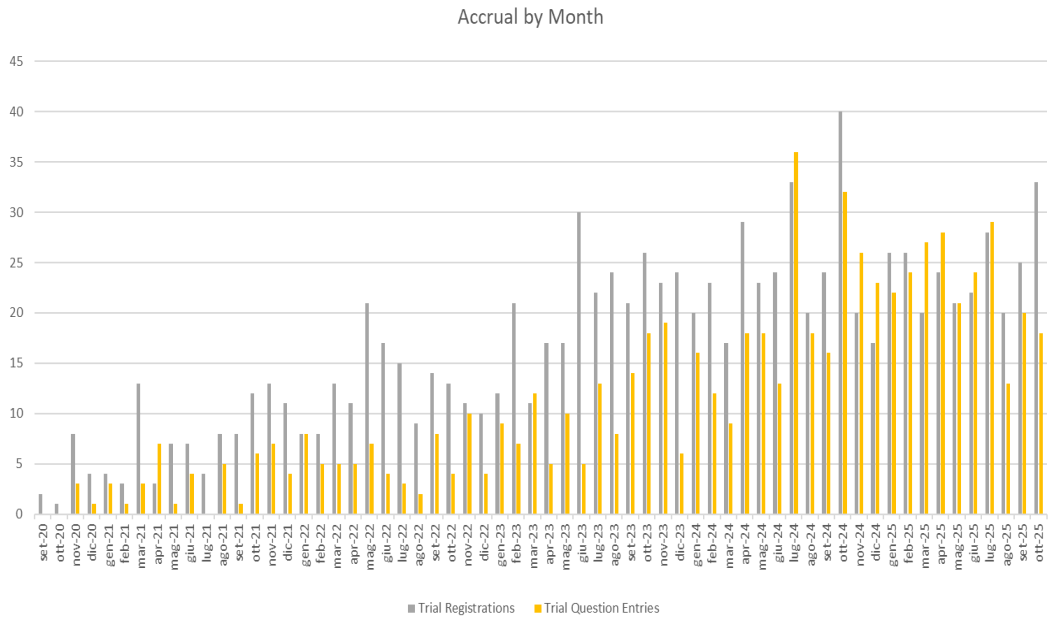
Studi aperti/in apertura

Attività EpSSG

«Globale»

INSTRUCT

FaR-RMS
Frontline and Relapse
RhabdoMyoSarcoma study



Aggiornamento Dicembre 2025

- **Aperte 20 nazioni**
- **155 Centri**
- **1044 pazienti registrati**
- **721 arruolamenti nei trial randomizzati**

(1 pz può essere arruolato in più trial)

- **26 novembre 2025 <u>ingresso Italia</u>**

Padova (NCC) – 3 pz

Contratti inviati ai Centri (AIEOP)

CRO contatterà i centri per le SIV

Torino in attesa SIV

FaR-RMS Treatment Questions Open

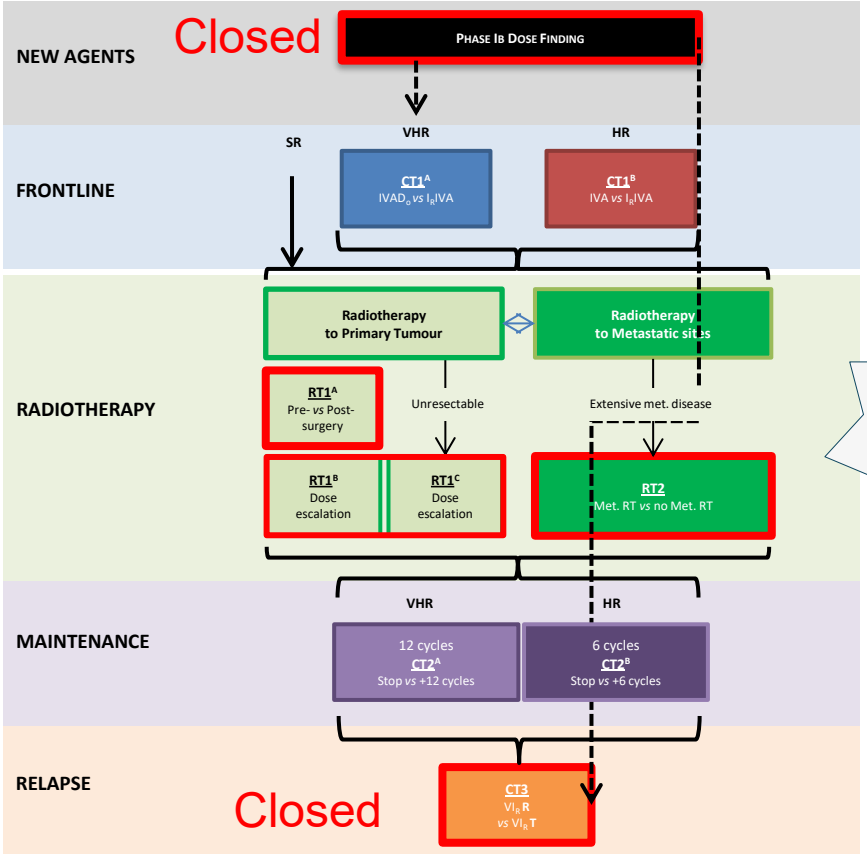
DW-MRI scans collected

PET-CT/ MRI scans collected

RT1a & RT2 Closed as of 21.07.2025

RT1b & RT1c Suspended as of 11.02.2026

Biological Sample Collection



Pathology samples at diagnosis

Surgical Reports Uploaded

Quality of Life RT1a and RT2

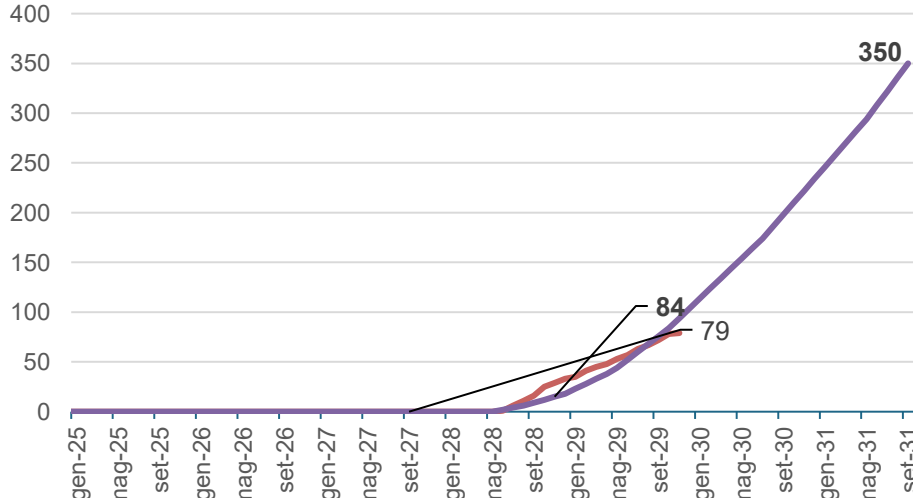
Quality of Life CT3

Upcoming Protocol Amendment

- Confirmation of closure of RT1a, RT2 and CT3
- Addition of wording to allow randomisation of patients with tumours with MYOD1 L122R mutation
- Addition of biological endpoints and changes to biology samples collection
- Additional quality of life questionnaire and change to collect quality of life from more patients
- Additional wording to ensure protocol is CTIS compliant

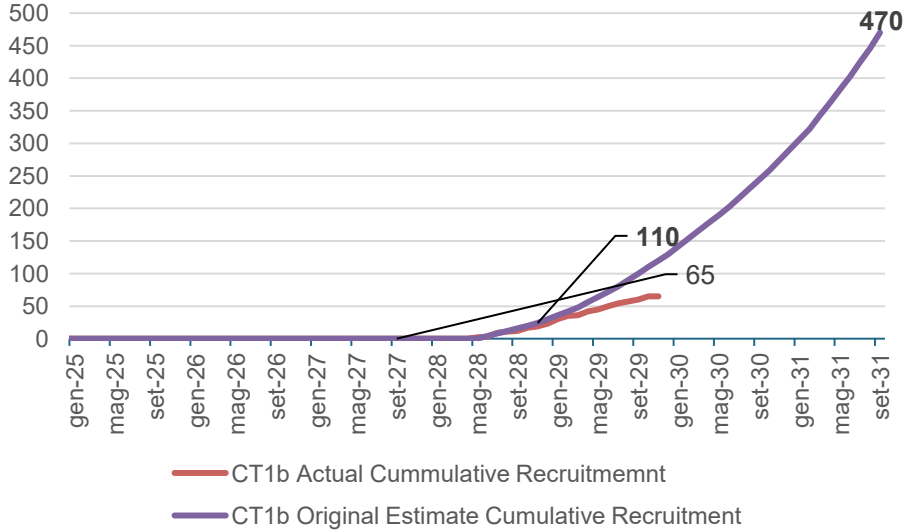
CT1a Recruitment

Very High Risk: Induction Chemotherapy

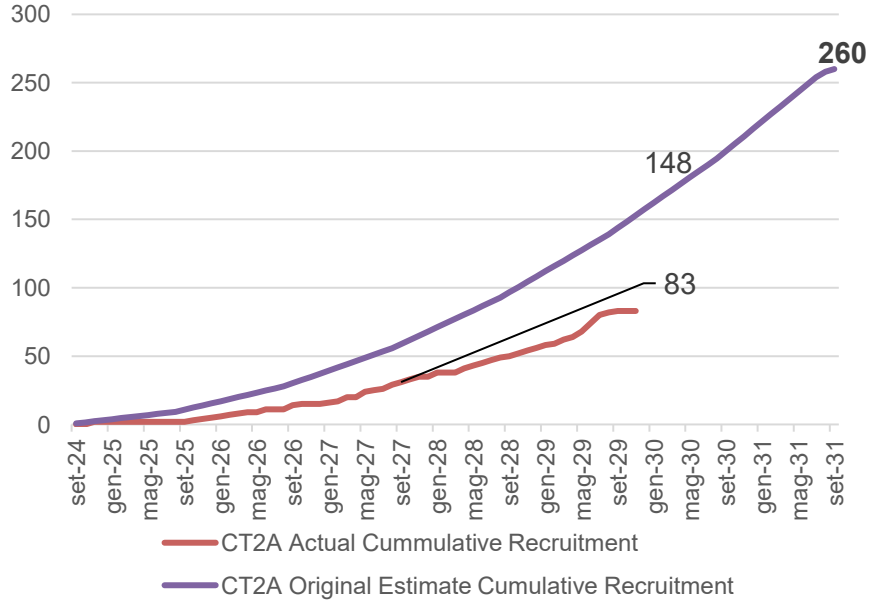


CT1b Recruitment

High Risk: Induction Chemotherapy



Data correct as of 05-Nov-2025

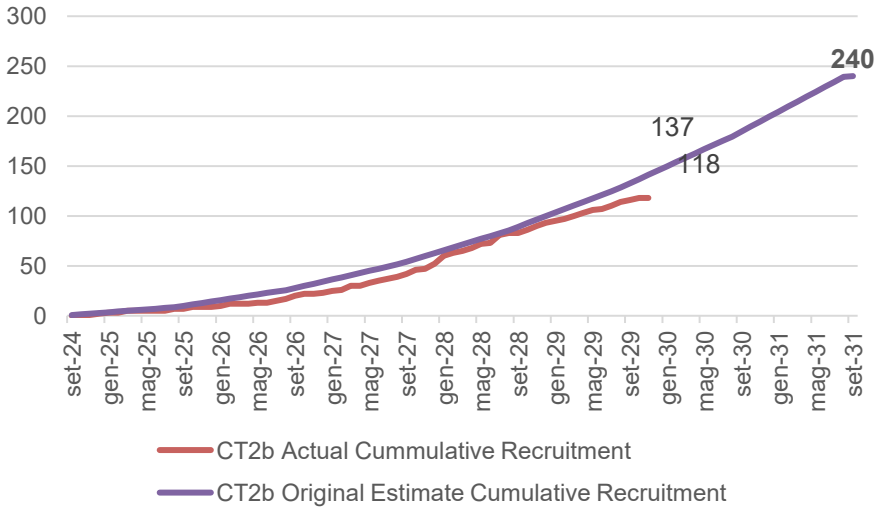


CT2a Recruitment

Very High Risk: Maintenance Chemotherapy 1 vs 2 yrs

CT2b Recruitment

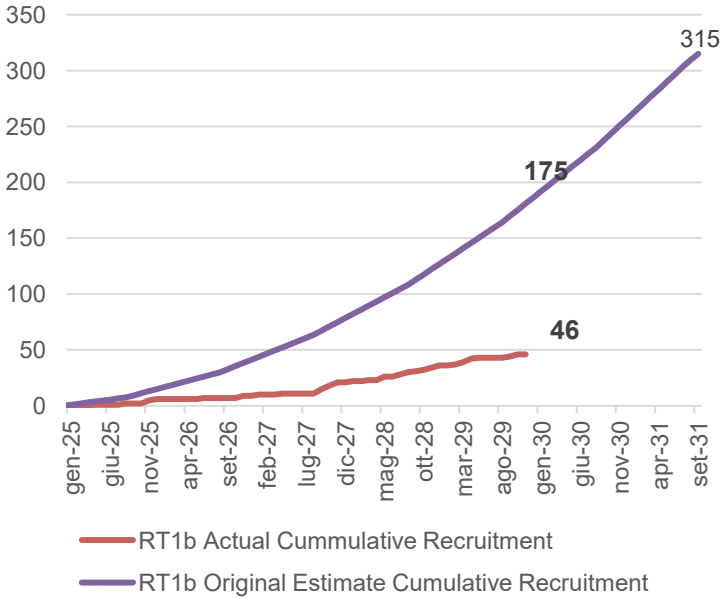
High Risk: Maintenance Chemotherapy: 6 no vs 1 yr



Data correct as of 05-Nov-2025

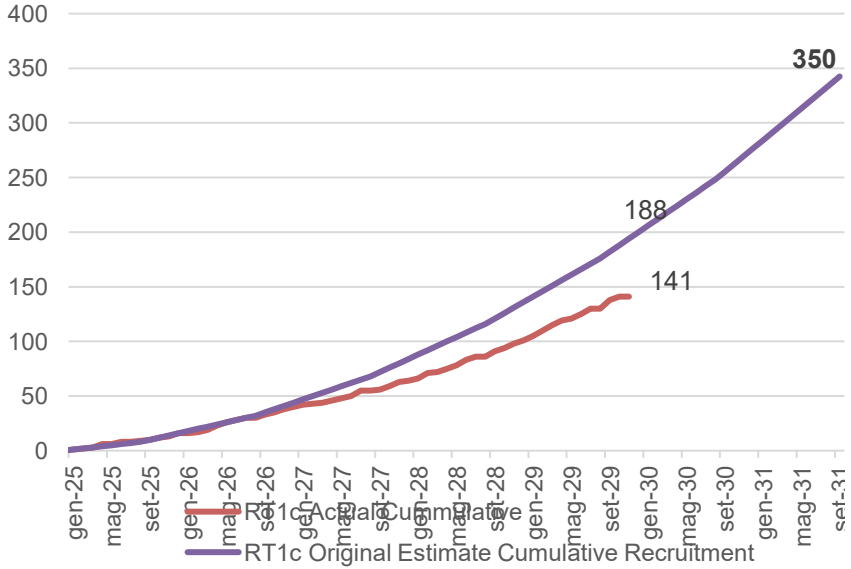
RT1b Recruitment

Dose escalation of RT in patients
with a higher local failure risk (resectable disease)



RT1c Recruitment

Dose escalation of RT in patients
with a higher local failure risk (unresectable disease)



Data correct as of 05-Nov-2025

RIMINI

Molecular Identification and Characterization of non-Rhabdomyosarcoma Soft Tissue Sarcoma in Kids, Adolescents and Young Adults: an E*p*SSG NRSTS study

MyKids is a non-interventional, observational, international, multicentre, prospective study with invasive measurements

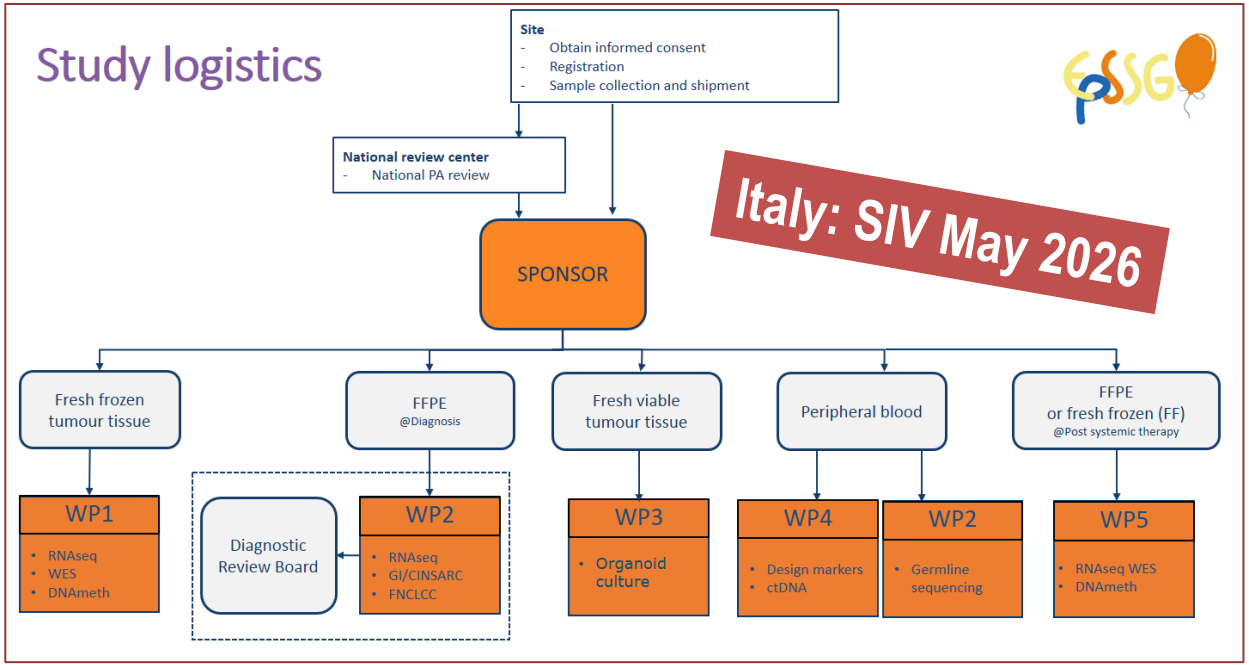
Aims

- The overall aim is to better understand the potential for molecular diagnosis of pediatric NRSTS with a view to optimize treatment.
- International pathology review (20% misdiagnosis historical cohort) = WP2
- Investigate prognosticators NRSTS (GI, CINSARC) = WP2
- Molecular diagnostics NRSTS = WP1&WP2
- Develop organoid models = WP3
- Develop and investigate liquid biopsies = WP4
- Identify post therapy clones = WP5

Inclusion Criteria:

- Suspected NRSTS or ≤2 months after diagnosis of NRSTS
- Age: 0-25 years
- Written informed consent

Minimal requirements: Diagnostic FFPE material and peripheral blood for germline analysis for participation in WP2.



OCTOPUS

Optimising Combination Therapy for Paediatric, Adolescent and Young Adult Patients with Non-Rhabdomyosarcoma Soft Tissue Sarcomas

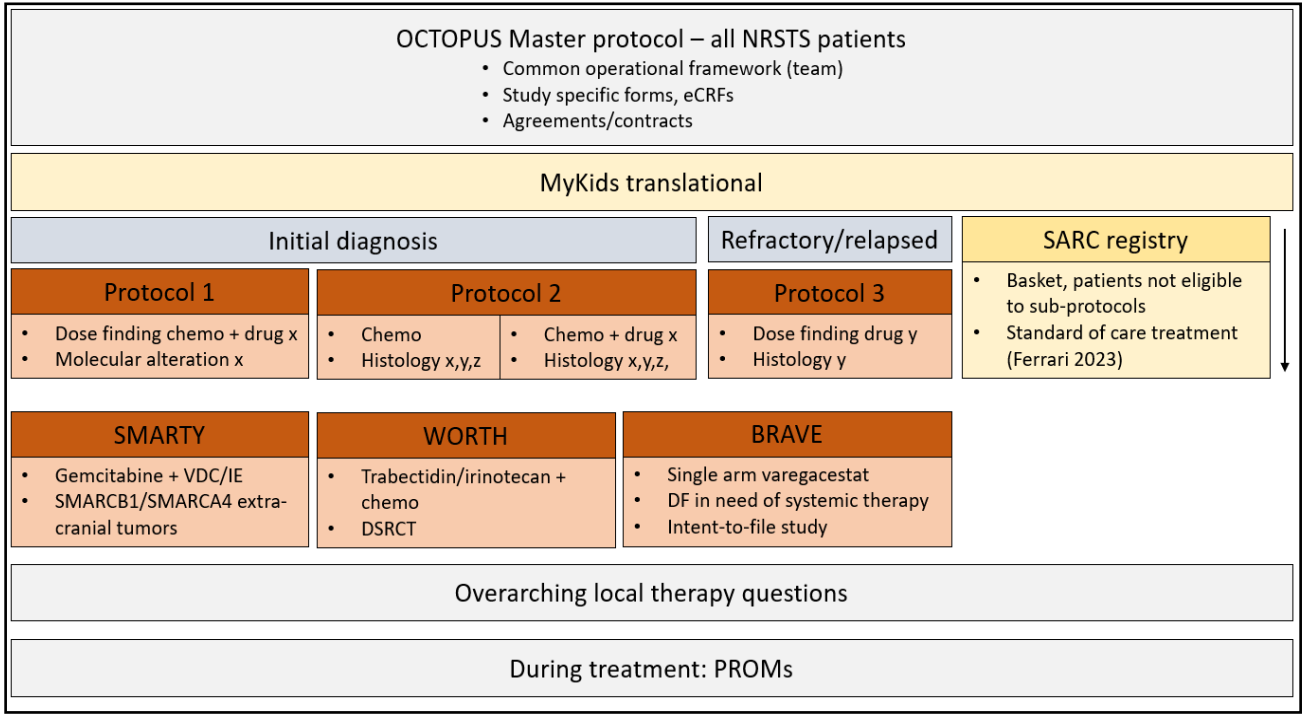
OCTOPUS is a master protocol for a platform trial that includes sub-trials and a SARC registry for children, adolescents and young adults with NRSTS, linked to the diagnostic E*p*SSG MyKids study.

OCTOPUS aims to accelerate the introduction of innovative treatments and to integrate translational research to deepen the understanding of these tumours.

Its adaptive design allows multiple sub-trials to run in parallel or consecutively, enabling faster, more efficient evaluation of new therapies. Patients not enrolled in sub-trials will receive standard-of-care treatment and will be registered in the SARC registry for real-world data collection.

OCTOPUS will provide a legal and operational framework to support trial development, data management, and organization of national coordinating centres and sites.

Overarching studies on local therapy, patient-reported outcomes, and translational science will be embedded within the platform.



OCTOPUS

Optimising Combination Therapy for Paediatric, Adolescent and Young Adult Patients with Non-Rhabdomyosarcoma Soft Tissue Sarcomas

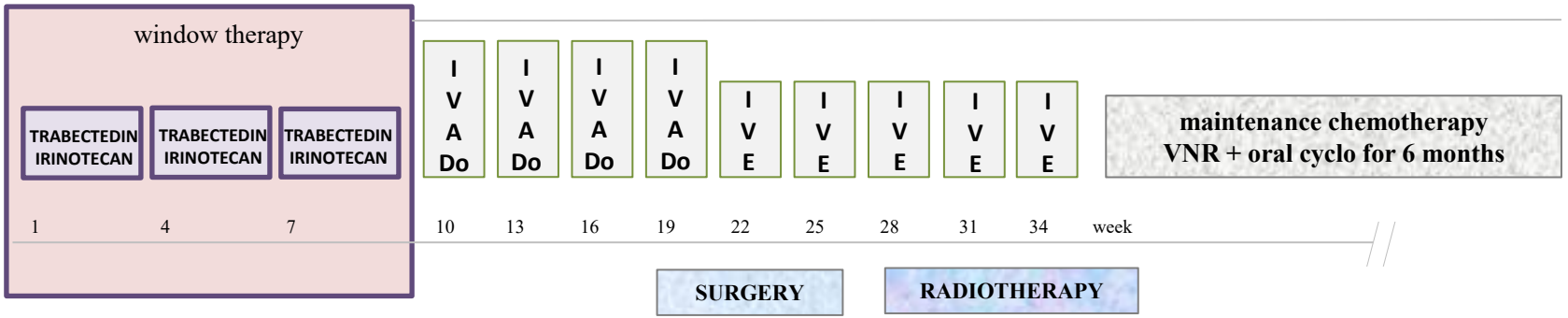
- Finance:
  - Start up grant obtained
  - Grant application for further financing pending
- First Studies:
  - 2 academic 'backbone' arms for frontline treatment
  - 1 pharma collaborations signed

- **WORTH.** (phase 1b) to determine the MTD/RP2D of the combination of trabectedin and irinotecan in a window phase, and (phase 2) to determine the activity of the combination of trabectedin and irinotecan in patients with newly diagnosed **desmoplastic small round cell tumours (DSRCT).**
- **SMARTY.** (phase 1b) dose confirmation study adding gemcitabine to conventional chemotherapy and in maintenance, and (phase 2) to determine the efficacy of combining gemcitabine with conventional VDC/IE chemotherapy and in maintenance in patients with newly diagnosed extra-cranial **malignant rhabdoid tumours** (ecMRT).
- **BRAVE.** multicentre, single arm, open-label, phase 2 study investigating the safety and efficacy of varegacestat, a selective gamma secretase inhibitor, in paediatric patients with newly diagnosed or relapsed/refractory **desmoid-type fibromatosis**. The study will be independently funded through a collaboration with Immunome

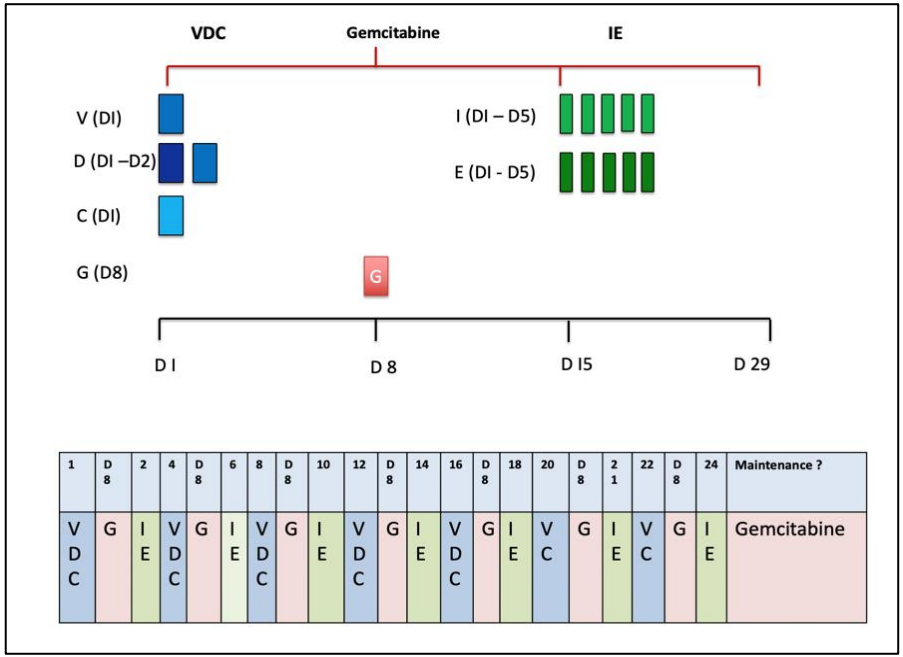
WORTH Study

Window Therapy with Irinotecan and Trabectedin in patients with Desmoplastic Small Round Cell Tumor

WORTH. (phase 1b) to determine the MTD/RP2D of the combination of trabectedin and irinotecan in a window phase, and (phase 2) to determine the activity of the combination of trabectedin and irinotecan in patients with newly diagnosed desmoplastic small round cell tumours (DSRCT).

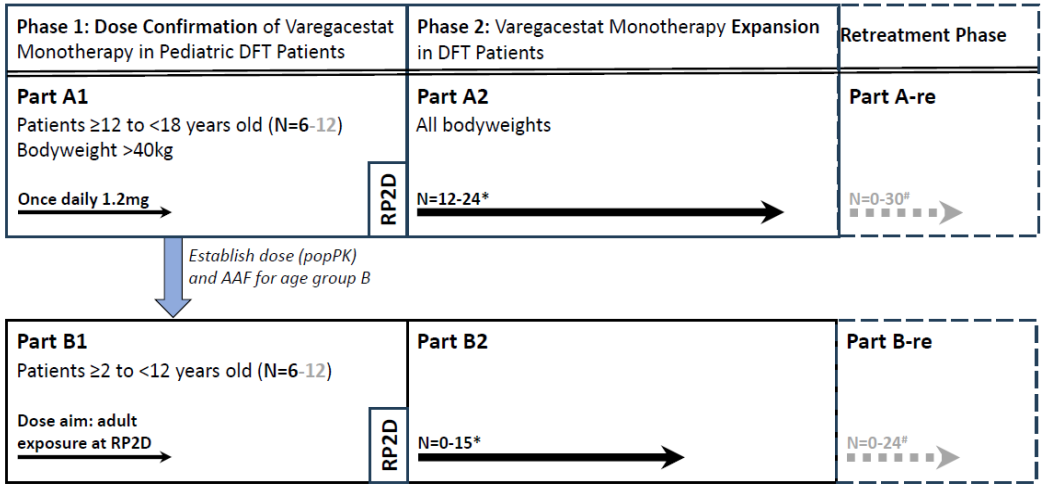


SMARTY. (phase 1b) dose confirmation study adding gemcitabine to conventional chemotherapy and in maintenance, and (phase 2) to determine the efficacy of **combining gemcitabine with conventional VDC/IE chemotherapy** and in maintenance in patients with newly diagnosed **extra-cranial malignant rhabdoid tumours (ecMRT).**



1	D 8	2	4	D 8	6	8	D 8	10	12	D 8	14	16	D 8	18	20	D 8	2 1	22	D 8	24	Maintenance ?
VDC	G	IE	VDC	G	IE	VDC	G	IE	VDC	G	IE	VDC	G	IE	VC	G	IE	VC	G	IE	Gemcitabine

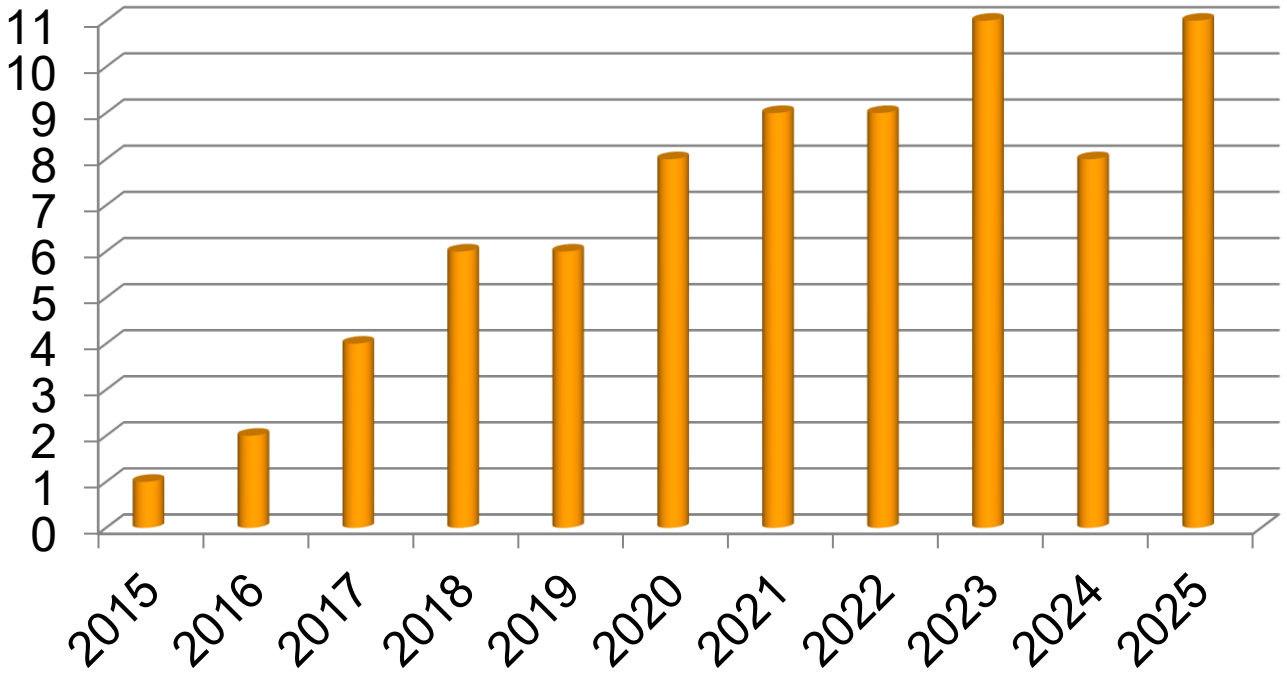
BRAVE. multicentre, single arm, open-label, phase 2 study investigating the safety and efficacy of **varegacestat, a selective gamma secretase inhibitor**, in paediatric patients with **newly diagnosed or relapsed/refractory desmoid-type fibromatosis.** The study will be independently funded through a collaboration with Immunome



* For **Part B**: minimum N=6 treated at RP2D (during **B1** and **B2** combined), maximum N=18. In total (Part A & B) N=36 treated at RP2D.
# All subjects enrolled in this trial may re-enter the Retreatment Phase after stopping initial treatment, if meeting retreatment in-/exclusion criteria
This design requires 3-12 additional subjects depending on timing of DLTs + dose we end up with as RP2D (0-6 in Part A & 3-6 in Part B)

EpSSG Activities update

Publications by year



- 75 publications total
- 131 EpSSG members with at least 1 publication

EpSSG Ongoing analyses and New proposal

-29 ongoing studies: (9 Italian P.I. +5 italian colleagues as collaborators)

- 8 new studies after RMS WG-calls
- 6 new proposal

-8 ongoing studies with other groups (COG, former CWS) - 1 Italian P.I.

Beatrice, Ilaria e Julia

	INSTRuCT Studies				
1	**Addendum 4 Biology/pathology group FOXO1 vs PAX3 vs PAX7**	S. Hettmer	04/06/2024	01/09/2024	**Report v6 sent 30 May. Presentation at the meeting 3 June. Abstract for CTOS sent, accepted as oral ppt. Draft sent 1-July, IDC sent Report v.7 and new tables 13 AUG. Waiting for treatment data to be uploaded. Next meeting Nov**
2	Cerebrospinal fluid diagnostics in patients with parameningeal RMS EpSSG-COG	A. Weis, R. Knops, R. Schoot, JHM	06/07/2022		**Report with EpSSG data to be prepared and shared with A.Weiss, then reconsider the proposal**
3	INSTRuCT and Oberlin 2.0	JHM Merks, Chisholm	20/01/2025		Proposal sent to the IDC 20.01.2025. JHM to contact Suzi for the data delivery. **Addendum to be signed in Padova, then start with the analyses (IDC will see the data first, then kickoff meeting). High priority. Data received December, report to be**
4	Relapsed localized RMS	J. Chisholm	03/02/2026		**proposal submitted Feb 3, 2026; JC to contact BC in March, draft by June 2026**
5	INSTRuCT risk stratification analysis	EpSSG-COG	18/11/2021		**discussed during INSTRuCT meeting Paris December 2024 and June 2025**
	Other Studies				
1	Leiomyosarcoma CWS/EpSSG series	CWS-EpSSG-	15/06/2021		**write to AF for update_ to do with Monika Sparber Sauer (After Body Size)**
2	WLI	CWS-EpSSG	15/09/2024		IDC sent EpSSG data to CWS people 26 April 2024-**paper circulated November 3rd**
3	Paraspinal RMS proposal	P. Soloni	17/12/2024	11/09/2024	Proposal sent 17.12.2024, JHM to review and share with the Board, IDC sent list of pts June, sample size to be defined, waiting for CWS data (sent reports of EpSSG 11.09) plan kick-off. **CWS data received Sep 30th, dataset integrated. Data pending, analyses will**

2. Analysis by primary tumor site: Candidates & project (09.02.2026)

1. Nasal Ala and Nasolabial Fold (33 pts): Working Group: **Ida RUSSO**, GM Milano, JHM Merks

- **Authors list:** Ida Russo, Giuseppe Maria Milano, Sofia Rahman, Marinka Hol, Raquel Davila Fajardo, Hans Merks, Rita Alaggio, Veronique Minard, Daniel Orbach, Julia Chisholm, Henry Mandeville, Gabriela Guillén Burrieza, Andrea Ferrari, Gianni Bisogno.

2. Airway RMS (oral cavity, oropharynx, larynx, trachea, and hypopharynx: 40 pts)

Working Group: **Marinka HOL,** W. Breunis, G. Bisogno

3. Local Control Approaches and Their Impact on Survival in Thoracic (39 pts) and trunk (20 pts) RMS: Working Group: **Federico MERCOLINI**, F. De Corti, A. Kelsey

4. Intraoperative challenges and postoperative outcomes of patients with pelvic RMS (81 pts)

Working Group: **Sarah BRAUNGART,** T. Rogers, F. Guerin

5. Abdominal (24 pts) and abdominal wall (7 pts) RMS Working Group: **Anna CAMPELLO, Antonio RIBELLES, Trung NGUYEN,** J. Chisholm

1. Rare entities project update (25.03.2026)

VGLL-NCOA2: Patrizia Gasparini, *Jessica Morgan, Rita Alaggio*, Monika Sparber-Sauer

- registered in the RAYYAN platform and in PROSPERO. 1st draft by the end of March (update 9.02)
- Some difficulties in involving people in papers selection

TP53 fusion: Francois Sevrin, Joanna Selfe, Janet Shipley, Julia Chisholm, Veronique Minard-Colin:

-first draft to be circulated in March (update 9.02)

-abstract submitted to SIOP2026; (update 25.03)

EWSR1/FUS:TFCP2: Amadeus Heinz, Silvia Pomella, Rossella Rota, Michael Meister, Hans Merks:

-Dataset is mature, Biology section manuscript is ready (update 9.02)

Wednesday 3rd Dec 2025 - 16.00-17.30

RMS closed session

«Towards the next EpSSG protocol».

Chair: G. Bisogno

Agenda

- 16.00-16.10 Welcome and RMS WG (G.Bisogno)
- 16.10-16.25 MEK-inhibitors for the frontline treatment of RMS_ an update (W. Breunis & M. Meister)
- 16.25-16.40 Possible 'new' agents of interests toward new trial. (W. Breunis)
- 16.40-16.55 CaR-T cell news (M. Sparber-Sauer)
- 16.55-17.10 Maintenance Chemotherapy (LL. Hjalgrim)
- 17.10-17.20 ADC available for RMS patients – LIGHTBEAM protocol (M. Casanova)
- 17.15-17.30 Open discussion (All)

Prossimo meeting
Glasgow 2026

The EpSSG hammer is back in Italy !!

Associazione Italiana Ematologia Oncologia Pediatrica

STSC Soft Tissue Sarcoma Committee

Nazionale

Riunioni GDL

Studi aperti/in apertura

Pubblicazioni

Europeo

Studi aperti/in apertura

Attività EpSSG

«Globale»

INSTRUCT

Il Consorzio INSTRuCT

Consorzio: INternational Soft Tissue SaRcoma ConsorTium (INSTRuCT).

Fondazione: Istituito a Copenhagen nel 2017 da gruppi Nordamericani (COG) ed Europei (CWS, SIOP-MMT, ICG ed EpSSG).

Missione: Creare un **set di dati comune** per superare la rarità della patologia e facilitare lo **scambio** di informazioni.

Supporto: Supervisionato dal Pediatric Cancer Data Commons (PCDC).

Sviluppo di **accordi di condivisione dati** tra i diversi sponsor degli studi e l'Università di Chicago.

Il Problema e la Necessità di Armonizzazione

Attualmente

COG (Nord America)

gruppi europei

COG e i **gruppi europei** utilizzano sistemi di stratificazione del rischio differenti.

Sistemi discordanti

Le differenze nei criteri di rischio ostacolano il confronto tra i risultati dei vari studi clinici internazionali.

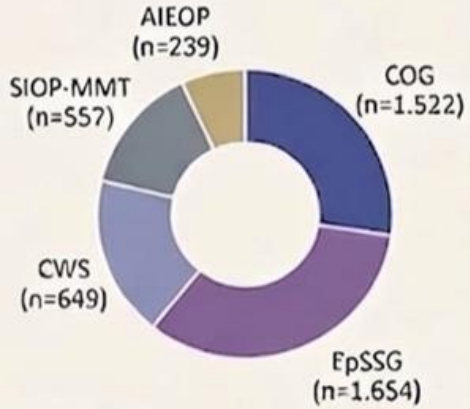
Obiettivo

Sviluppare un sistema di **stratificazione del rischio armonizzato** e applicabile a livello internazionale utilizzando dati aggregati.

Caratteristiche della Coorte Analitica

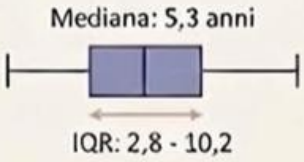
Totale Pazienti: **4.621**

Gruppi Cooperativi



Dati Demografici

Età alla Diagnosi



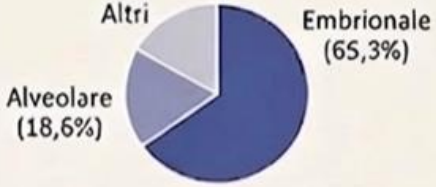
Sesso

Maschi: 61%

Femmine: 39%

Caratteristiche del Tumore

Istologia



Sedi Primarie Comuni

GUnonBP

HNPM

Dimensioni

≤5 cm: 52,9%

Stadiazione e Stato Molecolare

Stadio T

N0

N0: 85,5%

Gruppo IRS

Gruppo III: 69,6%

Stato di Fusione FOXO1

Negativo: 86,0% (3.974)

Positivi alla Fusione

PAX3::FOXO1: 63,7% (412/647)

Risultati: Modello di Cox per EFS

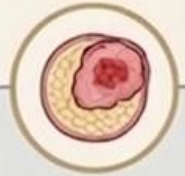
Età

- <3
  Ref (0.5 Punti)
- 3 to 12
  HR 0.74 [0.65;0.83]
  (0 Punti) 0
- >12
  HR 1.01 [0.86;1.17]
  p=0.9234
  (0.5 Punti) 0.5

Sede

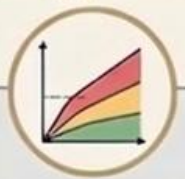
- GUnonBP
  Ref (0 Punti) 0
- GUBP/Orbita
  HR 1.18 [0.93;1.50]
  p=0.1733
  (0 Punti) 0
- HNnoPM
  HR 1.59 [1.23;2.05]
  p<0.001
  (1 Punto) 1
- Estremità/HNPM/Altro
  HR 1.66 [1.33;2.07]
  p<0.001
  (1 Punto) 1

T



- T1
  Ref (0 Punti) 0
- T2
  HR 1.33 [1.18;1.49]
  p<0.001
  (0.5 Punti) 0.5

Gruppo IRS



- Gruppo I
  Ref (0 Punti) 0
- Gruppo II
  HR 1.3 [1.00;1.69]
  p=0.0535
  (0.5 Punti) 0.5
- Gruppo III
  HR 1.75 [1.37;2.25]
  p<0.001
  (1 Punto) 1

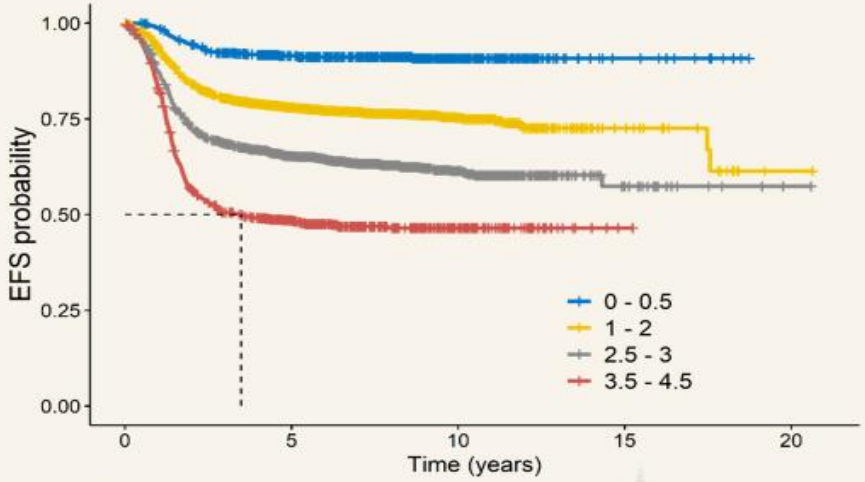
Dimensione

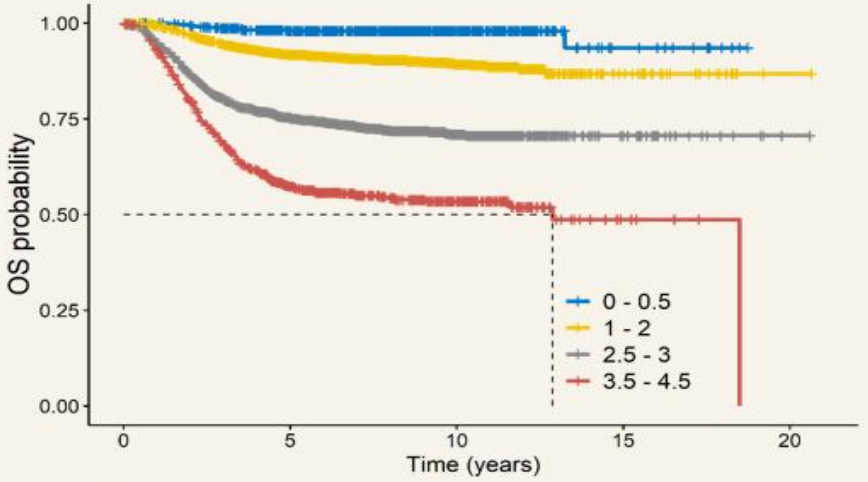
- <=5 cm
  Ref (0 Punti) 0
- >5 cm
  HR 1.35 [1.20;1.51]
  p<0.001
  (0.5 Punti) 0.5

Fusion status

- Assente
  Ref (0 Punti) 0
- Presente
  HR 1.61 [1.40;1.84]
  p<0.001
  (1 Punto) 1

Le Nuove Categorie di Rischio





At risk

528	391	136	13	0
1739	1047	293	22	1
1622	838	240	17	1
732	265	64	1	0

At risk

528	422	147	14	0
1739	1252	379	30	1
1622	970	277	24	1
732	315	86	6	0

		Low (0-0.5) (N=528)	Standard (1-2) (N=1739)	Intermediate (2.5-3) (N=1622)	High (3.5-4.5) (N=732)
5yr EFS	% (95%CI)	91.4% (88.7;93.6)	78.0% (76.0;79.9)	65.3% (62.9;67.6)	48.5% (44.8;52.2)
5yr OS	% (95%CI)	98.0% (96.3;98.9)	91.7% (90.3;92.9)	75.3% (73.1;77.3)	57.3% (53.5;60.9)

48 projects

- **10 Approved by the designated Committee, writing not yet underway**
- **15 in progress: data has been released, or writing is underway**
- **3 Presented: project has been presented at a conference but not yet published**
- **20 published**

INSTRuCT Research

Click on a description to view the original project proposal.
(ST) = statistician

roject Status Key
pproved: Project has been approved by the designated committee; writing not yet underway.
Progress: Data has been released or writing is underway.
esented: Project has been presented at a conference but not yet published.
ublished: Paper has been published.

INSTRuCT #	Principal Investigator	Description	roject Type	Status	Publication	Presentation	COG Investigators
2026-02	Julia Chisholm	First relapse/progression of localized RMS: an analysis of clinical features and outcomes from the International Soft Tissue saRcoma ConsorTium	Investigator	Approved			
2025-07	Douglas Harrison Leo Mascarenhas *Weo Xue (ST)*	Outcomes of Adolescents and Young Adults (AYA) with Rhabdomyosarcoma	Investigator	Approved			Douglas Harrison Leo Mascarenhas Weo Xue (ST)
2025-06	Federico Mercolini Gianni Bisogno	Impact of local treatments on non-lung single site metastatic patients: an INSTRuCT Analysis	Investigator	Approved			
2025-05	Sarah Winter *Matthieu Carton (ST)*	The International Soft Tissue Sarcoma Consortium: The baseline analysis of Non-rhabdomyosarcoma soft tissue data	WG Research	Approved			Aaron Weiss Navid Fariba Jonathan Metts
2025-04	Anne-Sophie Defachelles *Marie-Cécile Le Deley (ST)*	Clinicobiological Characterization of Pediatric Clear Cell Sarcoma of Soft Tissue Using the INSTRuCT Database	Investigator	Approved		"	Jacquelyn Crane Timothy Lautz
2025-02	Doug Hawkins	THE INTERNATIONAL SOFT TISSUE SARCOMA CONSORTIUM (INSTRUCT) AS A MODEL FOR DATA SHARING AND CONSENSUS DEVELOPMENT	WG Consensus	Presented		SIOP 2025 - Poster walk	
2025-01	Hans Merks *Beatrice Coppadora (ST)*	Reevaluation of Oberlin Score including FOXO1 status	Investigator	In Progress			Nathan Schloemer Jamie Aye Wendy Allen-Rhoades Dana Casey
2024-01	Gianni Bisogno	The INternational Soft Tissue SaRcoma ConsorTium (INSTRuCT): the baseline analysis of rhabdomyosarcoma data	WG Research	Published	The International Soft Tissue Sarcoma Consortium: The baseline analysis of rhabdomyosarcoma data DOI: 10.1002/cncr.35974	SIOP 2025 - ePoster	Dave Rodeberg Suzanne Wolden Raj Venkatramani Sam Volchen boum Doug Hawkins
2023-01	Tim Lautz *Jim Anderson (ST)*	Correlation of clinical and pathologic lymph node staging in RMS of the extremity	WG Research	In Progress			Tim Lautz Thomas Scharschmidt Roshni Dasgupta Dave Rodeberg

https://commons.cri.uchicago.edu/instruct/

Prossimi appuntamenti

			2026
7 May (Thu)	SIOPE Congress (EpSSG Spring meeting)	**Glasgow**	Confirmed
December 2-4 (Wed-Fri)	EpSSG Winter Meeting & Association Assembly	**Padova**	Confirmed
			2027
April or May	SIOPE Congress (EpSSG Spring meeting)	**TBD**	
December 1-3 (Wed-Fri)	EpSSG Winter Meeting & Association Assembly	**Stuttgart**	Confirmed

CEVAIE

J Clin Oncol. 2004 Dec 1;22(23):4787-94.

European intergroup studies (MMT4-89 and MMT4-91) on childhood metastatic rhabdomyosarcoma: final results and analysis of prognostic factors

M Carli [1], R Colombatti, O Oberlin, G Bisogno, J Treuner, E Koscielniak, G Tridello, A Garaventa, R Pinkerton, M Stevens

IVADo

The **IVADo** regimen--a pilot study with ifosfamide, vincristine, actinomycin D, and doxorubicin in children with metastatic soft tissue sarcoma: a pilot study of behalf of the European pediatric Soft tissue sarcoma Study Group.

Bisogno G, Ferrari A, Bergeron C, Scagnellato A, Prete A, Alaggio R, Casanova M, D'Angelo P, Di Cataldo A, Carli M.

Cancer. 2005 Apr 15;103(8):1719-24. doi: 10.1002/cncr.20928.

Addition of dose-intensified doxorubicin to standard chemotherapy for rhabdomyosarcoma (**EpSSG RMS 2005**): a multicentre, open-label, randomised controlled, phase 3 trial.

Bisogno G, Jenney M, Bergeron C, Gallego Melcón S, Ferrari A, Oberlin O, Carli M, Stevens M, Kelsey A, De Paoli A, Gaze MN, Martelli H, Devalck C, Merks JH, Ben-Arush M, Glosli H, Chisholm J, Orbach D, Minard-Colin V, De Salvo GL; European paediatric Soft tissue sarcoma Study Group.

Lancet Oncol. 2018 Aug;19(8):1061-1071. doi: 10.1016/S1470-2045(18)30337-1. Epub 2018 Jun 22.

IrIVA

Integrating irinotecan in standard chemotherapy: A novel dose-density combination for high-risk pediatric sarcomas.

Bisogno G, Ferrari A, Tagarelli A, Sorbara S, Chiaravalli S, Poli E, Scarzello G, De Corti F, Casanova M, Affinita MC.

Pediatr Blood Cancer. 2021 Jul;68(7):e28951. doi: 10.1002/pbc.28951. Epub 2021 Mar 10.

Vinorelbina/Ciclofosfamide

Vinorelbine and low-dose **cyclophosphamide** in the treatment of pediatric sarcomas: pilot study for the upcoming European Rhabdomyosarcoma Protocol.

Casanova M, Ferrari A, Bisogno G, Merks JH, De Salvo GL, Meazza C, Tettoni K, Provenzi M, Mazzarino I, Carli M.

Cancer. 2004 Oct 1;101(7):1664-71. doi: 10.1002/cncr.20544.

Vinorelbine and continuous low-dose **cyclophosphamide** as maintenance chemotherapy in patients with high-risk rhabdomyosarcoma (RMS 2005): a multicentre, open-label, randomised, phase 3 trial.

Bisogno G, De Salvo GL, Bergeron C, Gallego Melcón S, Merks JH, Kelsey A, Martelli H, Minard-Colin V, Orbach D, Glosli H, Chisholm J, Casanova M, Zanetti I, Devalck C, Ben-Arush M, Mudry P, Ferman S, Jenney M, Ferrari A; European paediatric Soft tissue sarcoma Study Group.

Lancet Oncol. 2019 Nov;20(11):1566-1575. doi: 10.1016/S1470-2045(19)30617-5. Epub 2019 Sep

FaR-RMS
Frontline and Relapse
RhabdoMyoSarcoma study

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- Ida Russo
- Luisa Santoro
- Pietro Soloni
- Stefania Cardellicchio
- ……

Team Padova
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- Angela Scagnellato
- Alessandra Paratella
- Francesca Cassaro
- Debora Vanuzzo

……

CENTRI AIEOP

Il Giardino della Ricerca
Per la ricerca e la cura di sarcomi e tumori rari pediatrici