AIEOP
ASSOCIAZIONE ITALIANA EMATOLOGIA ONCOLOGIA PEDIATRICA

Presidente
A. Mastronuzzi

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

Giornate AIEOP

GDL Tumori del sistema nervoso centrale

Maura Massimino

Sistema Socio Sanitario
Fondazione IRCCS Istituto Nazionale dei Tumori
Regione Lombardia

RIMINI
Hotel Savoia

13-14 aprile 2026

Disclosures of Name Surname

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

Giornate AIEOP

RIMINI 13-14 APRILE 2026
















TC e Riunioni recenti

- **Riunione open del GdL 25 Febbraio 2026**
- **BTG SIOPE Porto settembre 2025**
- **Retreat HGG Graz ottobre 2025**
- **Riunione SIOPe HGG 2.3 marzo 2026**
- **Aggiornamenti relativi ai protocolli aperti/in fieri**
- **Plurime consulenze/discussioni casi per mail/telefoniche/*de visu***
- ***proposte***

Programma odierno

- Aggiornamento protocolli aperti
  - Medulloblastoma
    - Dati acquisiti
    - Protocollo HR
  - Ependimoma
  - «Registro», prot. Osservazionale DIPG
  - Legge 648 giugno 2025 (DMG)
  - Gliomi a basso grado: LOGGIC, protocollo in corso
  - Tumori teratoidi/rabdoidi atipici

- Protocolli (ancora) in apertura
  - Medulloblastoma < 3 aa alla diagnosi
  - Quasi al traguardo protocollo HGG SIOPe
  - Epilogue per basso grado

- Protocollo osservazionale tumori cerebrali AIEOP
- Proposte di studio

Significato della centralizzazione dei tumori pediatrici CNS

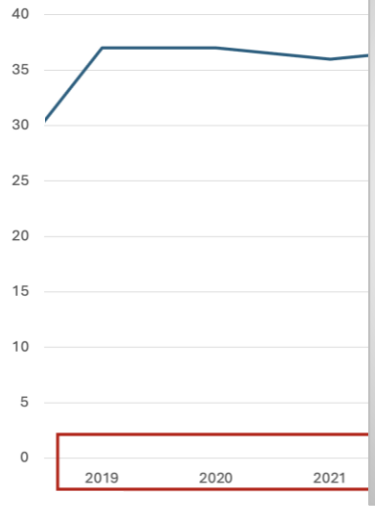
alla Neuropatologia di Sapienza Università di Roma

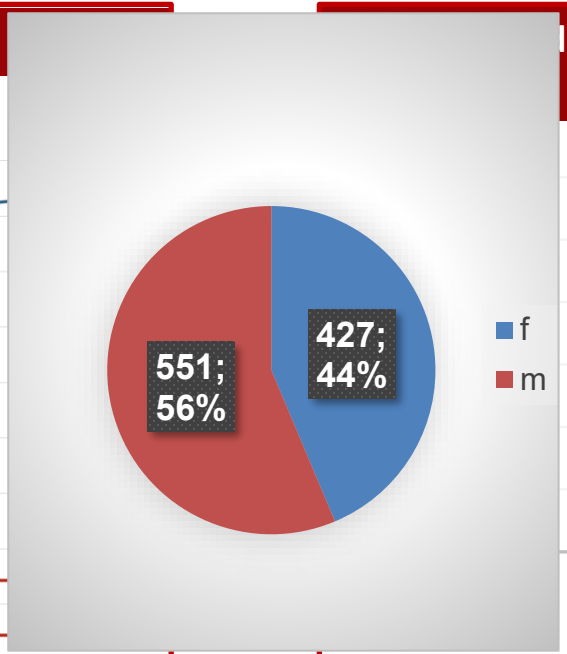
- ✓ Valutazione consultiva dei tumori del SNC, **Attività di consulenza** per colleghi specialisti in casi diagnostici difficili
- ✓ **Correlazione diretta** tra dati molecolari, morfologia e istopatologia, con revisione esperta centralizzata.
- ✓ **Equità nazionale di accesso** alle analisi molecolari mirate a supporto della diagnosi, indipendentemente dalla sede geografica.
- ✓ **Assistenza a Formazione** continua nella diagnostica neuropatologica dei tumori, Formazione di giovani collaboratori nei metodi moderni di neuropatologia tumorale
- ✓ **Supporto** a progetti scientifici e studi terapeutici neuro-oncologici in qualità di centro di riferimento
- ✓ Sviluppo e valutazione di nuove procedure diagnostiche di patologia molecolare, **Implementazione rapida** di analisi genetico-molecolari.
- ✓ **Standardizzazione e riproducibilità:** uniformità dei protocolli analitici, riduzione dell'impatto della variabilità pre-analitiche.
- ✓ **C**

Miglioramento dell'accuratezza diagnostica con riduzione dei casi borderline e maggiore precisione nella classificazione molecolare, migliore stratificazione prognostica ed accesso a protocolli terapeutici mirati.

ANDAMENTO CENTRALIZZAZIONE ISTOLOGICA 2023-2025

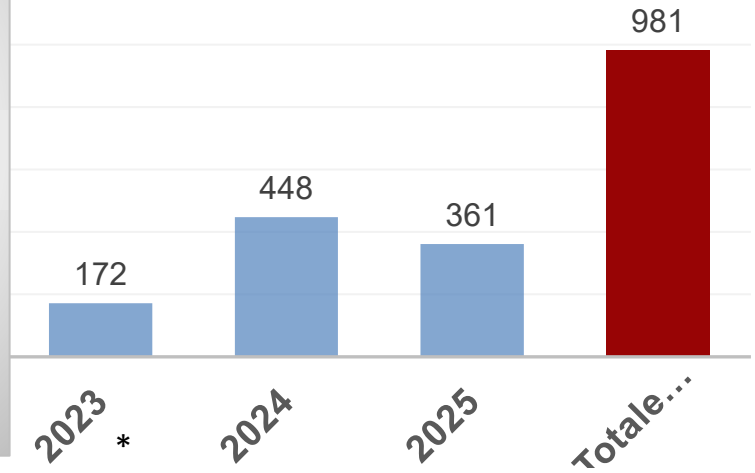
GdL 2023





2019: 449 — 37 / mese
2020: 452 — 37 / mese
2021: 434 — 36 / mese
2022: 442 — 37 / mese

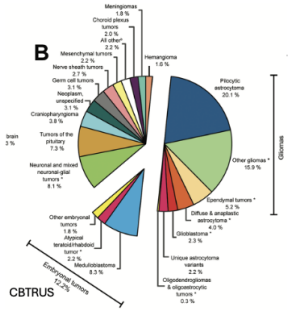
li osservazione agosto 2023- dicembre 2025
Totale 981



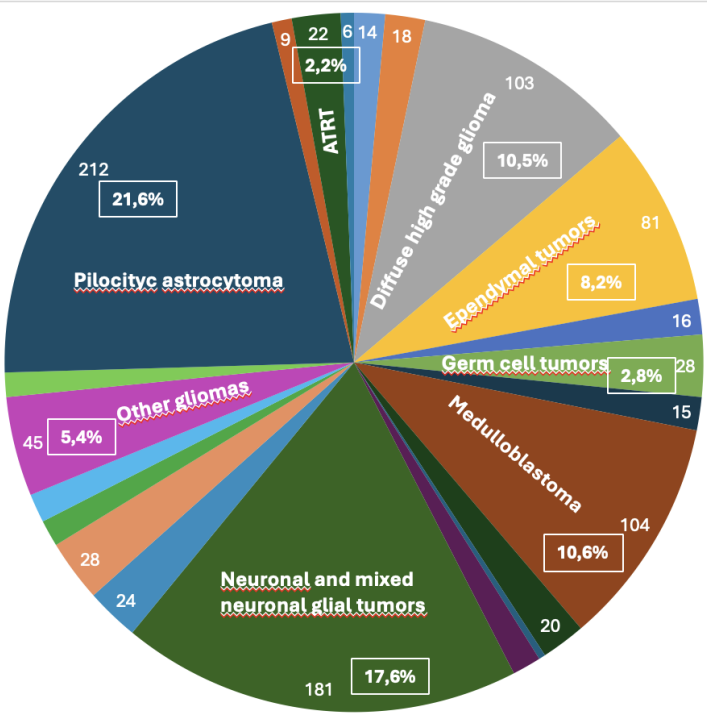
* Dati anno 2023
mesi agosto – dicembre (5 mesi)

172/5= 34.4 / mese

Età media 10,5 (range 0-25 yrs)



CBTRUS



Choroid plexus tumors	1,4%	14
Craniopharyngioma	1,8%	18
Diffuse high grade glioma	10,5%	103
Ependymal tumors	8,2%	81
Focal cortical dysplasia	1,6%%	16
Germ cells tumors	2,8%%	28
Materiale esiguo per diagnosi	1,5%	15
Medulloblastoma	10,6%	104
Meningeal tumors	2,0%	20
Mesenchymal tumors	0,3%	3
Nerve sheat tumors	1,3%	13
Neuronal and mixed neuronal-glial tumors	17,6%	181
No tumors / non diagnostic	2,4%	24
Non neoplastic patholgy	2,8%	28
Oligo-astrocityc tumors IDH mutated	1,2%	12
Other embryonal tumors	1,3%	13
Other glioma	5,4%	45
Other tumors	1,1%	11
Pilocytic astrocytoma	21,6%	212
Pituitary tumors	0,9%	9
Tumore teratoide rabdoide atipico (ATRT)	2,2%	22
Tumors of the pituitary	0,6%	6

Total 981

44% LGG
AP
Glioneuronal
Gliomas

Dati agosto 2023 dicembre 2025

Gene	Molecular alteration	Methods
MGMT promoter	methylated/unmethylated	MS-PCR
IDH1/2 mutations	mutations ex. 4 (cod 132, 172)	Sanger ex.4 (+ IHC R132H), ddPCR, NGS
1p/19q	codeletion	FISH + MLPA
1q	gain	FISH
6q	deletion	FISH
CMYC	amplification	FISH + ddPCR
NMYC	amplification	FISH + ddPCR
chr 6	monosomy	FISH + ddPCR
CTNNB1 (β-cat)	mutations ex. 3 (cod 32,33,34	Sanger ex.3 (+ IHC β-cat), NGS

Numbers of molecular centralization CNS pediatric tumors: last **9** years

	2017	2018	2019	2020	2021	2022	2023	2024	2025
Molecular Analysis	189	207	203	COVID	241	363	415	596	655

2017: 84 FISH + 10 Seq + 80 PCR

2018: 116 FISH+ 26 Seq + 65 PCR

2019: 96 FISH + 36 Seq + 71 PCR

2021: 142 FISH + 36 Seq + 63 PCR

Mostly MYC/MYCN ampl, 1p/19q del, 6q, CDKN2A del, IDH1/2 mut

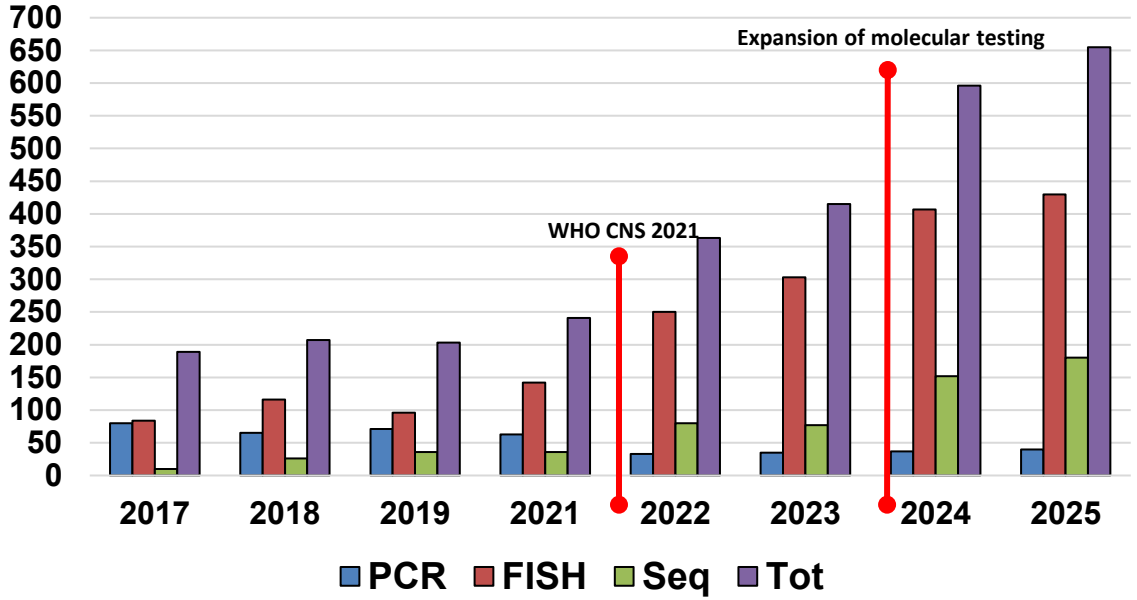
2022: 250 FISH + 80 Seq + 33 RT/digital PCR

2023: 303 FISH + 77 Seq + 35 PCR

2024: 407 FISH + 152 Seq + 37 RT/digital PCR

2025: 430 FISH + 180 Seq + 40 RT/digital PCR

Progressive integration of multiple analysis (C19MC, EGFR, CDKN2A, PDGFRA, H3F3A and other mut, fusions and rearrangments)



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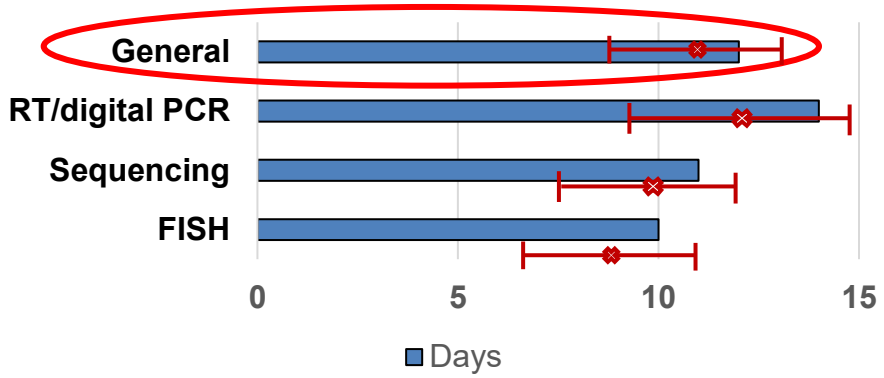
6q	deletion	FISH
CMYC	amplification	FISH + ddPCR
NMYC	amplification	FISH + ddPCR
chr 6	monosomy	FISH + ddPCR
CTNNB1 (β-cat)	mutations ex. 3 (cod 32,33,34 ..)	Sanger ex.3 (+ IHC β-cat), NGS
PTCH, SMO, SUFU, P53	mutations all exons	NGS
19q13MC (C19MC)	amplification	FISH (+ IHC Lin28)
BRAF	mutations ex15 (V600)	Sanger ex.15 (+ IHC V600E), ddPCR, NGS
TERT promoter	promoter mutations (C225T,	Sanger, ddPCR, NGS
DICER1	mutations ex24 e 25	Sanger, ddPCR, NGS
EGFR	amplification	FISH
chr +7/-10	gain chr 7 / loss chr 10	FISH
CDKN2A (p16)	homozigous deletion	FISH, ddPCR
H3F3A/B (H3.3)	K27/G34 mutations ex. 2	IHC + Sanger, NGS
KIAA1549/BRAF	fusion genes	FISH, ddPCR
C11ORF95 (ZFTA)	break	FISH
YAP1	break	FISH
FGFR1	break	FISH
FGFR2	break	FISH
MYB	break	FISH
MYBL1	break	FISH
BCOR	tandem duplication	FISH
PRKC-SLC44A1	fusion genes	FISH
MN1	break	FISH
PLAG1	break	FISH
PGFRA	amplification	FISH

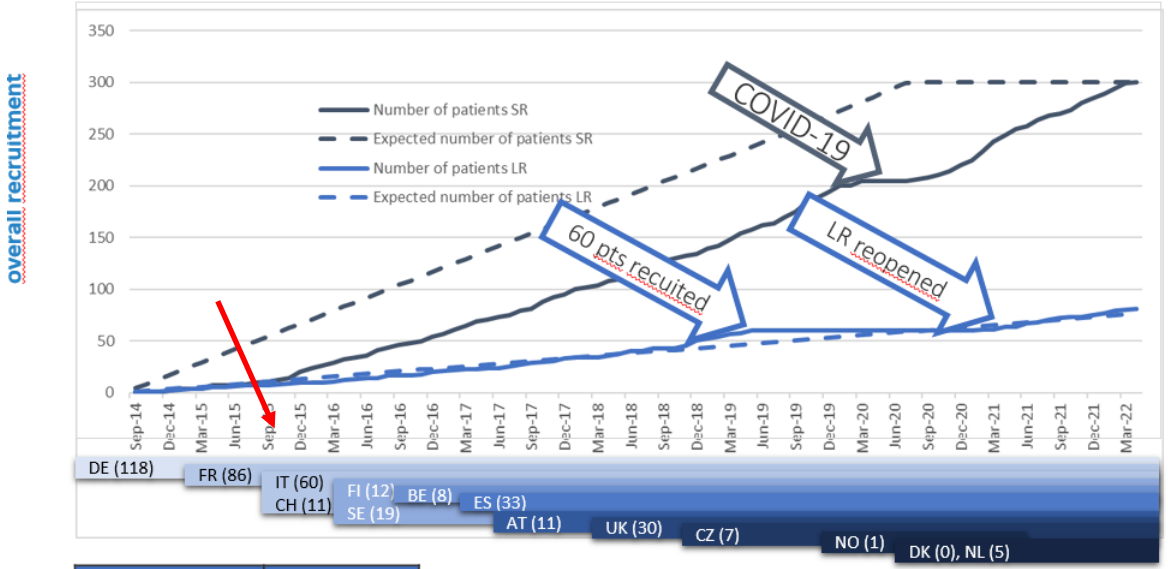
2025-26	n° Analysis	FISH	Sequencing (Sanger/NGS)	Rt/droplet digital PCR	Methylation
Molecular Analysis	655	430	180	40	Selected (OPBG)

TAT	Days Average	Days St.Dv
FISH	10	4
Sequencing	11	4
Rt/digital PCR	14	5
General	12	4



PNET 5 e MB6

PNET 5 – recruitment complete since March 2022



PNET 5 MB	Final status 04/2022
LR	81
SR	300
WNT-HR	11
SHH-TP53	6
Registry	3
all	401

Italia:
50 SR
10 LR

PNET 5 –

MB stu

P-PNET5-

astoma

PNET5 ri	
Low-	sion ional standard)
Standa	sion nal standard)
WNT-Hi (WNT with an	clinical trials
SHH-TP53 (any clinica	considered)

Re: Treatment of patients below 16 years of age presenting with R0M0 WNT group medulloblastoma

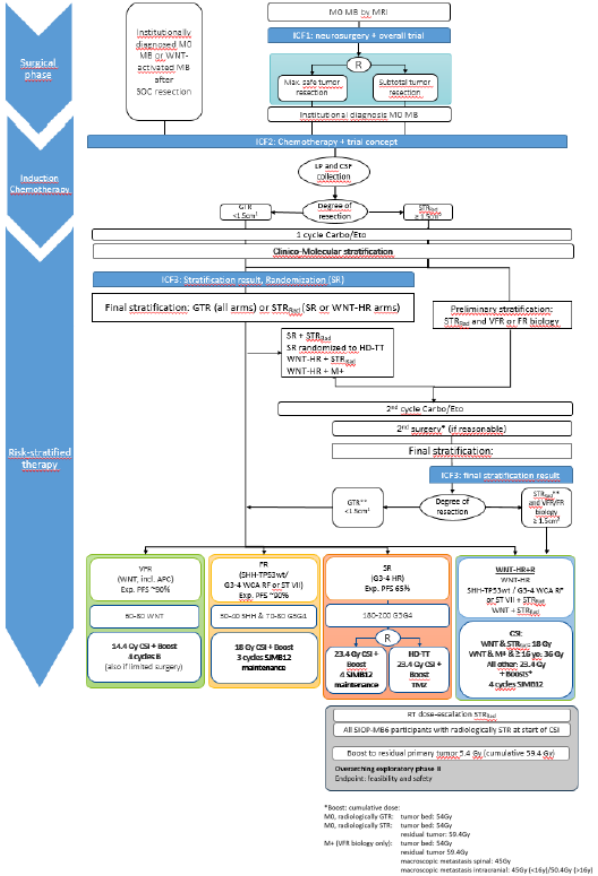
As you know, the first results of the PNET5 MB LR arm have been reported at the last SIOP meeting in Amsterdam, October 22, 2025 (See attached abstract).

The main objective was reached with a 3-year EFS rate of 91% in the per-protocol analysis (and 88.8% in the intent to treat analysis).

Based on these good results, and before the opening of the future SIOPE MB6 study, we may recommend today the **PNET5 MB LR treatment as a standard in R0M0 WNT group medulloblastoma diagnosed before 16 years** (as we agreed during our recent meeting in Porto).

However, this is only true in the conditions of the per-protocol treatment:

- Age <16.0 years at diagnosis
- Confirmed R0 (R<1.5 cm^2)
- Confirmed M0
- Classic or desmoplastic histology
- Confirmed WNT group: *CTNNB1* mutation AND at least one additional molecular feature of WNT medulloblastoma [defined as isolated monosomy 6 and/or WNT methylation group by DNA methylation or RNA expression profiling, using methods accredited according to national requirements], and no other biological parameter incompatible with this diagnosis. Please note: positive nuclear expression of β-catenin is NOT considered a robust marker for the definition of WNT status.
- Radiotherapy to be started within 28 days from surgery (maximum 40 days)
    - Careful review of radiotherapy plans
    - Brain 18 Gy in 10 daily fractions of 1,8 Gy
    - Spine 18 Gy in 10 daily fractions of 1,8 Gy
    - Primary tumor boost 36 Gy in 20 daily fractions of 1,8 Gy (total dose to primary tumor 54 Gy in 30 daily fractions of 1,8 Gy
    - Safety margins according to the PNET5 MB protocol recommendations
- Maintenance chemotherapy: To be started 6 weeks after end of radiotherapy
    - 6 cycles BA_BA_BA
    - Regimen B: Cyclophosphamide (1000 mg/m^2 day 1, 2) Vincristine (1.5 mg/m^2 day 1)
    - Regimen A: Cisplatin (70 mg/m^2 day 1) CCNU (75 mg/m^2 day 1) Vincristine (1.5 mg/m^2 day 1, 8,15)
    - Recommendations of dose adaptation according to PNET5 MB LR should be followed



SIOP MB6

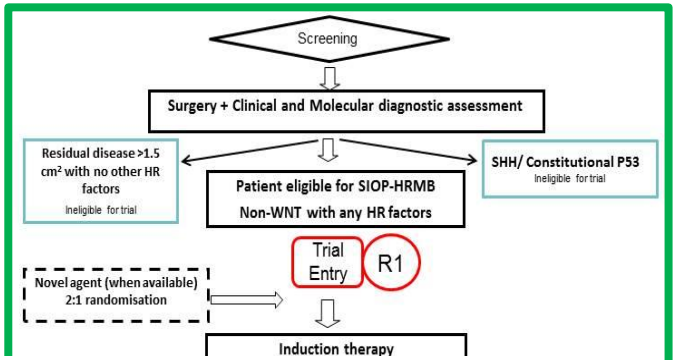
Dear Maura,
We have submitted the revised funding application in January 2026, it seems to be in review now and we expect to get a response in Q3/Q4 2026. If the response is positive we would aim at **a preparation in 2027**, with first wave of submissions and **trial site initiations in 2028**, and hopefully FPFV in Q4 2028. if all goes well.

Prof. Dr. med. Till Milde, MHBA

Total No patients	~380
Trial duration	6 (-8) years
Participating countries	SIOP-E
Sponsor	Germany

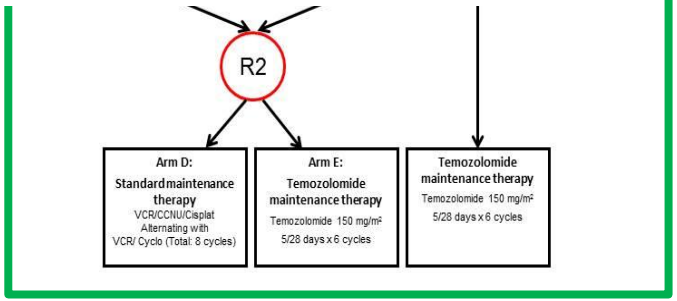
SIOP HRMB Trial

- Medulloblastoma (MB) is the most common malignant brain tumour in children and young people accounting for 20% of all brain tumours in children. Around 30% of MB patients are diagnosed as High-risk MB (HR-MB)
- Pha
- Chil
  with
  risk" medulloblastoma
- Screening phase
- 2 randomisations



Two hundred twenty-three patients have been screened on the trial:

- 107 patients have been randomised in Randomisation 1 (R1).
- 52 patients have been randomised in Randomisation 2 (R2)



Inclusion criteria – trial entry and R1

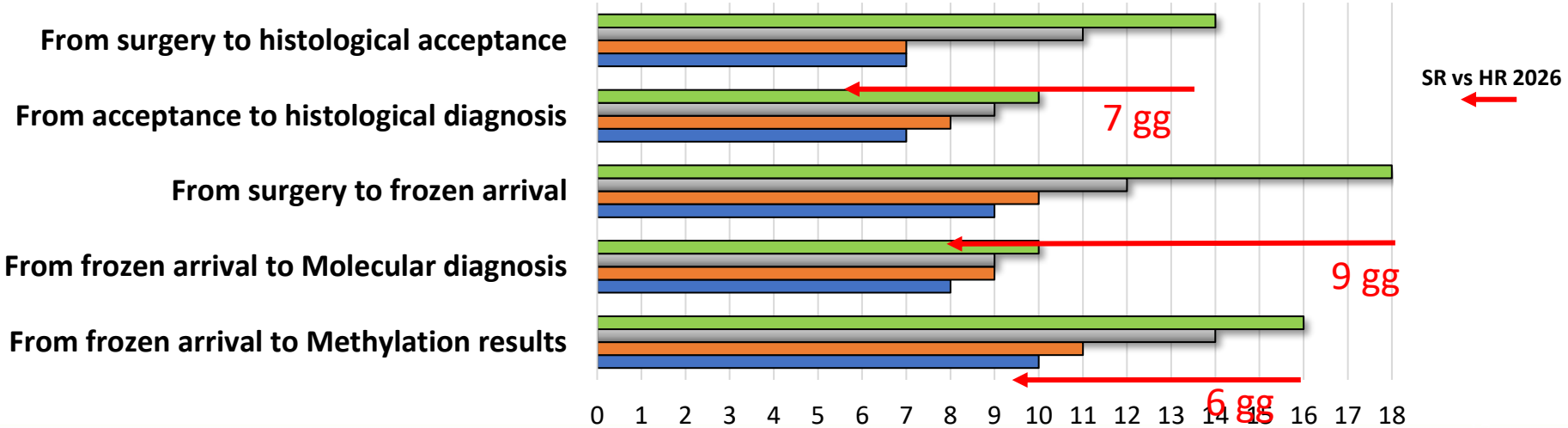
- Histologically proven (**centrally reviewed**) HR-MB with any of the currently defined histological subtypes
  - **SHH subgroup** or **non-SHH/non-WNT** (Groups 3 and 4)
  - With **at least one** of the following:
    - **Metastatic disease**: Chang stage M1, M2 and M3
    - **Large cell/Anaplastic MB** (defined by WHO criteria 2016)
    - **Significant residual tumour** (> 1.5 cm^2) following surgical resection of primary tumour and **other biological risk factors**
    - **Patients with MYC or MYCN amplified tumours** (unless MYCN amplified Group 4 without any other risk factors)
    - Patients with **SHH** subgroup tumours harbouring **somatic TP53 mutations**
- Age **≥ 3 years**
- **Submission of biological material**, including fresh frozen tumour samples and blood
- **No prior treatment for MB**, other than surgery, with the exception of one cycle of induction chemotherapy

	TAT PNET5-SR	
	Days Average	Days St.Dv
From surgery to histological acceptance	14	7
From histological acceptance to histological diagnosis	10	6
From surgery to frozen arrival	18	7
From frozen arrival to Molecular diagnosis	10	7
From frozen arrival to Methylation results	16	9

TAT HR-MB 2024	
Days Average	Days St.Dv
11	5
9	4
12	4
9	4
14	5

TAT HR-MB	
Days Average	Days St.Dv
7	3
8	5
10	4
9	3
11	5

TAT HR-MB 02/2026	
Days Average	Days St.Dv
7	2
7	4
9	5
8	3
10	5



Centralizzazione radioterapica, Quartet

QUARTET SITE APPROVALS/ICR

QUARTET RTQA Site Approvals

So far 35 institutions have completed the required site RTQA approval procedures, with a further 8 in progress. Site RTQA approvals are underway in Belgium, Czech Republic, Denmark, Finland, Germany, Italy, Netherlands, Norway, Sweden, and the United Kingdom.

ICR Case Review summary

* 60 cases reviewed (as of March 3rd)
* 28% (stable) required some kind of resubmission, mainly due to delineation
* Feedback is being acted upon by sites

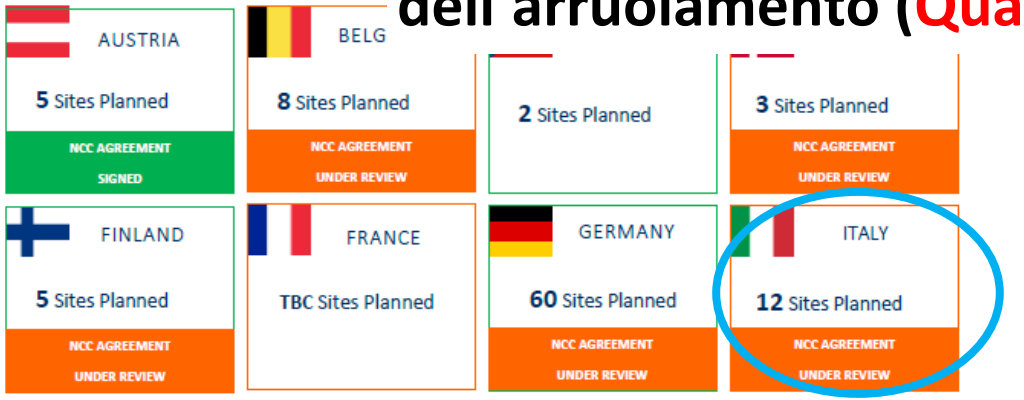
Siti Operativi:

- Milano
- Roma
- Napoli
- Genova
- Torino
- Udine/Aviano
- Bari
- Catania
- Padova
- Bologna

NATIONAL COORDINATING CENTRES SET-UP

The SIOP-HRMB trial is an international coll
the UK, we have 16 participating countries,

28 fe 2026 temporanea sospensione dell'arruolamento (Quartet business)



28 total patients

10/12 aperti

Udine 2 pz
Milano, 9 pz
Napoli, 8 pz
Roma BG, 8 pz
Genova, 1 pz

Protocollo osservazionale tumori embrionali rari, lavori in corso

Study aim

SIOP-E Registry for Patients with
Rare **E**mbryonal or **S**arcomatous CNS **T**umors

SIOP Europe
the European Society for Paediatric Onc

Gain further knowledge

by generating and analysing clinical, molecular, imaging and outcome data

Improve the stratification, treatment and outcome of patients

Develop and refine treatment recommendations

Develop hypotheses for prospective evaluation in subsequent trials

Rare embryonal tumours of the CNS:
- ETMR (embryonal tumour with multilayered rosettes)
- CNS neuroblastoma, FOXR2 activated
- CNS BCOR-altered tumours:
  - CNS tumour with BCOR internal tandem duplication
  - CNS tumour with BCOR(L1) fusion (provisional)
- CNS embryonal tumour NEC/NOS

Pineal tumours:
- Pineal Parenchymal Tumours:
  - Pineoblastoma
  - Pineal parenchymal tumour of intermediate differentiation (PPTID)
  - Pineocytoma
- Other pineal region tumours
  - Papillary tumour of the pineal region (PTPR)

Mesenchymal, non-meningothelial tumours of unknown differentiation, involving the CNS:
- CIC-rearranged sarcoma
- Primary intracranial sarcoma, DICER1-mutant
- Intracranial mesenchymal tumour, FET::CREB fusion-positive (provisional)
- CNS sarcomatous tumour NEC/NOS

Astroblastoma
- including MN1-altered and astroblastoma with other fusions

Recently described tumour types not included in the current WHO classification:
- CNS embryonal tumour with PLAGL-family amplification
- Neuroepithelial tumour with PATZ1 fusion
- CNS embryonal tumour with BRD4::LEUTX fusion

Patients with further molecularly defined tumour types, which may be identified within the spectrum of CNS-embryonal, CNS-sarcomatous tumours or other rare CNS tumour types not eligible for registration for other trial group trials/registries.

REST group meeting 25.09.25

Rare Embryonal and Sarcomatous CNS Tumour REST - Tumour Board

- Aim:
  - To provide expert advice for this rare group of CNS tumours for which there is generally no established standard of care and often poor prognosis
  - To provide some consistency / standardisation of care based on the evidence that is available and European guidelines
  - To offer inclusion in the SIOP-E CNS-REST Registry for patients discussed

- To meet every 2 weeks based on referrals
  - Currently alternating Tuesday 16.00-17.00 and Wednesday from 13.30-14.30 CET/CEST
  - 5 meetings since May; 12 patients discussed

CPMS 2.0 platform: https://cpms2.ern-net.eu/screen/home

CENTRE DE LUTTE CONTRE LE CANCER LEON BERARD

SIOP EPENDYMOMA II

An International Clinical Program for the diagnosis and treatment of children, adolescents and young adults with Ependymoma

EudraCT number: 2013-002766-39

Clinical Trials number: NCT02265770

VHP number: VHP201385

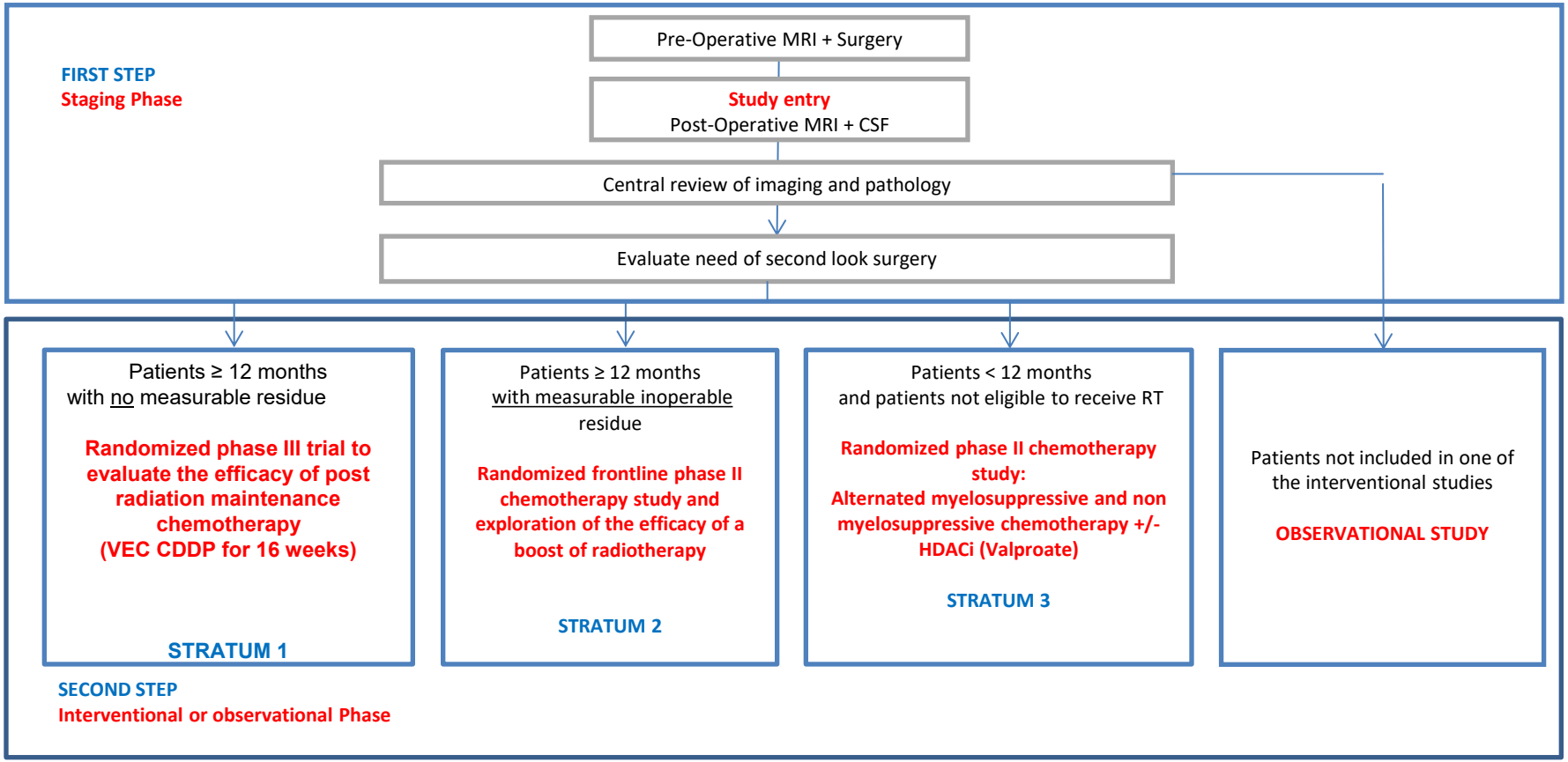
SOCIÉTÉ INTERNATIONALE D'ONCOLOGIE PÉDIATRIQUE
SIOP
INTERNATIONAL SOCIETY OF PAEDIATRIC ONCOLOGY

SIOP EP-II in Europe

15 Participating countries

Protocol version 4.0 dated 11/07/2022

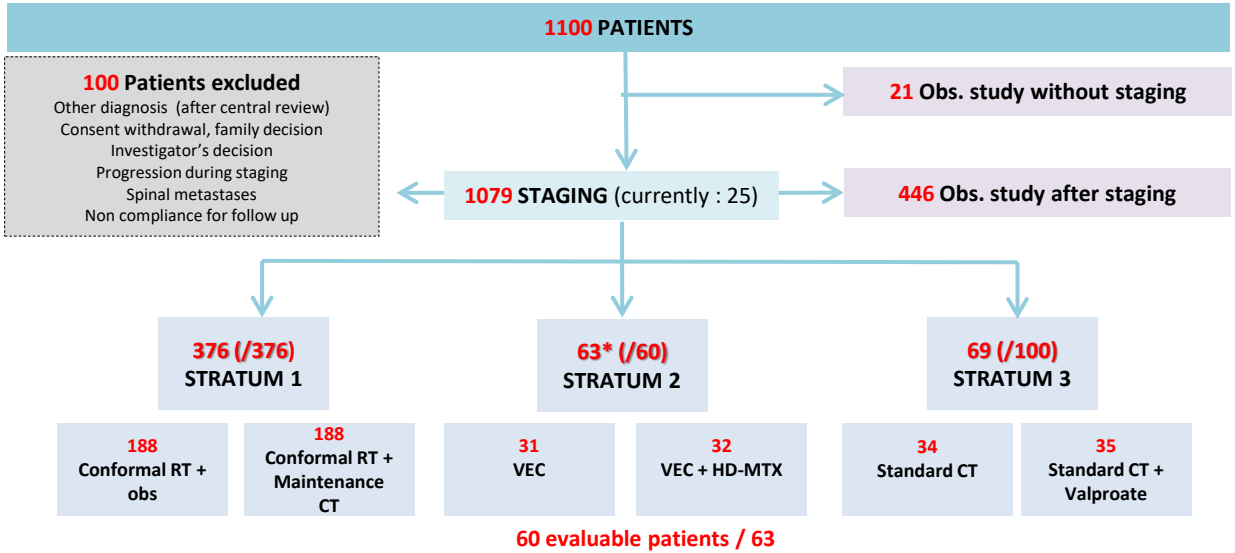
Study design



Overall recruitment

September 16th, 2025

EUROPE



60 evaluable patients / 63

All interventional strata are now closed to recruitment (observational open)

Overall recruitment Sept 16th, 2025	Activation date	Staging	Obs.	Stratum 1	Stratum 2	Stratum 3	Excluded	Total
France	April 2015	9	101	81	5	7	34	237
United Kingdom	December 2015	7	132	73	18	38	26	294
Italy	January 2016	0	44	92	10	2	12	160
Belgium	February 2016	0	22	12	4	2	5	45
Czech Republic	May 2017	0	7	8	0	5	0	20
Spain	October 2017	4	40	39	13	6	12	114
Ireland	November 2017	0	5	3	0	4	0	12
Austria	May 2018	0	3	10	0	0	1	14
Switzerland	May 2018	0	15	2	2	2	5	26
Finland	May 2018	0	3	1	0	0	0	4
Germany	August 2018	3	77	43	9	3	1	136
Netherlands	March 2020	0	12	5	1	0	4	22
Denmark	November 2020	1	2	2	1	0	0	6
Norway	December 2020	1	2	4	0	0	0	7
Greece	November 2021	0	2	1	0	0	0	3

IRCCS ISTITUTO NAZIONALE DEI TUMORI	**49** (100%)
A.O.U. OSPEDALI RIUNITI ANCONA	**5** (100%)
A.O.U. Integrata Verona	**4** (100%)
A.O.U. DI PARMA	**1** (100%)
A.O.U. SANTA MARIA DELLA MISERICORDIA, UDINE	**7** (100%)
IRCSS CRO DI AVIANO	**0** (0%)
AOU POLICLINICO VITTORIO EMANUELE CATANIA	**6** (100%)
AOU MEYER FIRENZE	**11** (100%)
AOB OSPEDALE ANTONIO CAO CAGLIARI	**0** (0%)
ISTITUTO GIANNINA GASLINI	**10** (100%)
OSPEDALE INFANTILE REGINA MARGHERITA	**14** (100%)
OSPEDALE PEDIATRICO BAMBINO GESÙ	**34** (100%)
HOSPITAL AORN SANTOBONO-PAUSILIPON, NAPOLI	**13** (100%)
Policlinico di Bari	**10** (100%)
A.O.U. BOLOGNA-S.ORSOLA-MALPIGHI	**3** (100%)

Upcoming final analysis (stratum 1)

- FA expected after 130 events (relapse or death)
- 106 events reported in the database (as of Sept. 17th 2025)
- 130th event expected November 2026

➢ **Please report any event of relapse or death in the database in real-time +++**

SIOP Europe
the European Society for Paediatric Oncology

SOCIÉTÉ INTERNATIONALE D'ONCOLOGIE PÉDIATRIQUE
SIOP
INTERNATIONAL SOCIETY OF PAEDIATRIC ONCOLOGY

What about future Stratum 1?

Maura Massimino

On behalf of ependymoma group

Phase 3 randomized trial of post-irradiation chemotherapy in patients with newly diagnosed ependymoma, a report from the Children's Oncology Group

(Neuro Oncol. 2025 Dec 19:noaf285. doi: 10.1093/neuonc/noaf285. Online ahead of print)

- (ACNS0831) Patients with PF (grade 2 or 3) or grade 3 ST ependymoma with GTR/NTR and patients with grade 2 ST ependymoma with GTR2 or NTR were randomized to 4 cycles of maintenance chemotherapy versus observation following RT
- Of 449 eligible patients, 325 with GTR/NTR or CR were randomized. Five-year EFS was 63.7% (95% CI 55.1-71.1%) for RT only (n=161) versus 69.2% (60.8- 76.3%) for RT-CHEMO (n=164) (one-sided log-rank p=0.299, HR=0.866). Fiveyear OS was 86.9% (79.8-91.6%) for RT only versus 88.3% (81.8-92.6%) for RTCHEMO (one-sided log-rank p-value=0.172, HR=0.757)
- **Primary analysis showed no benefit for maintenance chemotherapy**. Further follow-up is important to assess its effect on late relapses.

- **Patients ≥12 months without (relevant) residual disease (R0-R2):**
- Since the role of adjuvant chemotherapy is still unclear, patients ≥12 months without residual disease (R0-R2) should receive adjuvant focal radiotherapy only (total dose of 54-59.4 Gy in fractions of 1.8 Gy 5 times/week; total dose shall be adapted according to age, neurological condition and other risk factors such as multiple damaging surgeries or hydrocephalus).

What we may recommend in September 2025 for Stratum 2

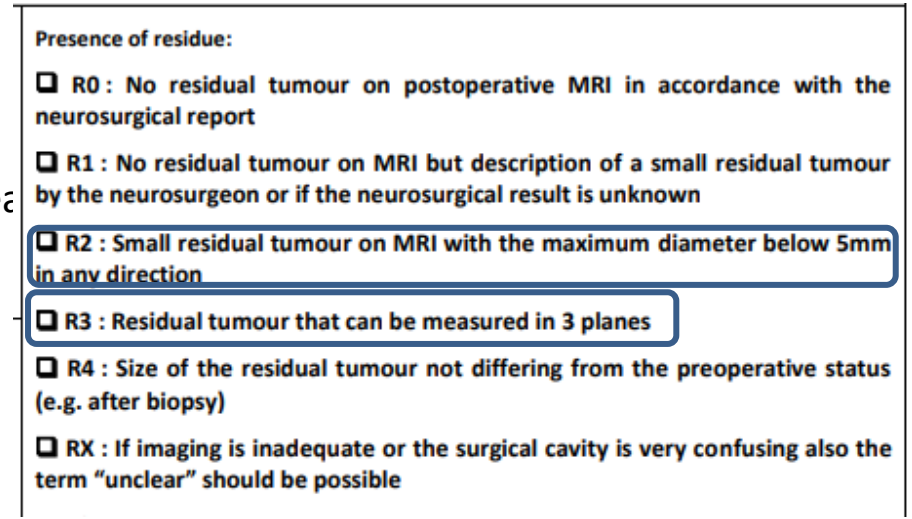
- **Patients ≥ 12 months with residual disease (R3-R4)**: **A second-look surgery** aiming at complete resection should always be evaluated both after first surgery and after pre-radiant chemotherapy. We recommend VEC courses (2-4) until random results available
- Post-operative radiation concepts may use boosts after standard dose according to SIOP II protocol in case of persistent measurable disease
- NEW… *consider second/further surgery after radiation time* (median time to response around 7 months in AIEOP series)

- *See also Obrecht D et al. Kinder und Jugendliche mit … Klin Padiatr 2023; 235: 167–177 | © 2023. Thieme.*

Ependimoma Stratum 2 Italia

- Valutazione della storia radiologica dei pazienti con Ependimoma arruolati nello STRATUM II in Italia dal 2016 al 2024

- 10 patients (1 N.A.)
- 9 patients : 4 females, 5 males
- Mean age at onset 5, 8 years (range 1-14 yea
- II look in 7/9



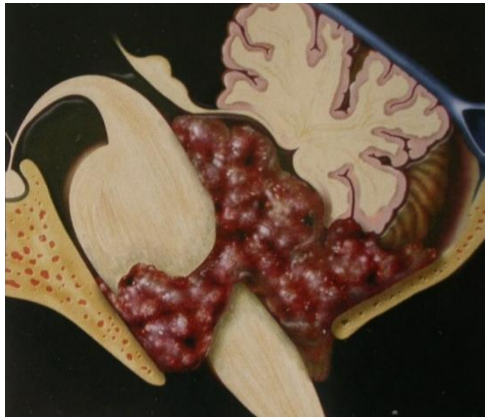
Presence of residue:

- ☐ R0 : No residual tumour on postoperative MRI in accordance with the neurosurgical report
- ☐ R1 : No residual tumour on MRI but description of a small residual tumour by the neurosurgeon or if the neurosurgical result is unknown
- ☐ R2 : Small residual tumour on MRI with the maximum diameter below 5mm in any direction
- ☐ R3 : Residual tumour that can be measured in 3 planes
- ☐ R4 : Size of the residual tumour not differing from the preoperative status (e.g. after biopsy)
- ☐ RX : If imaging is inadequate or the surgical cavity is very confusing also the term "unclear" should be possible

Results

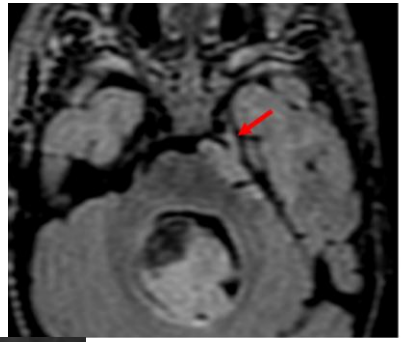
Pz. number	Post-surgery	Post- II III look	Post-chemotherapy
Pz 1	R3	R3	CR
Pz 2	R4	R3	SD
Pz 3	R3	R3	SD
Pz 4	R3	R3	SD, PR
Pz 5	R3	R3	PD
Pz 6	R3	-	SD, PR
Pz 7	R3	-	SD, PR
Pz 8	R3	R3	PR
Pz 9	R3	R3	SD

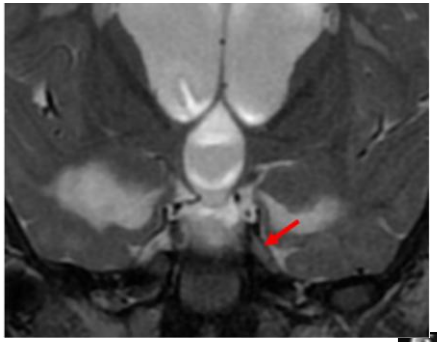
SD Stable disease; PR partial response; PD Progression disease; CR complete response



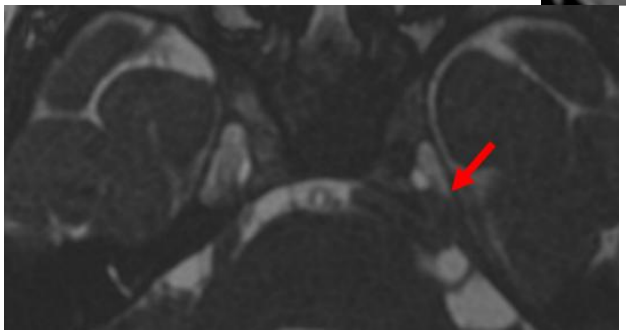
PF EPENDYMOMA

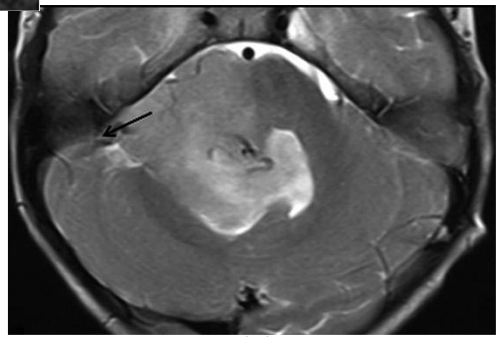
usefulness of 3D T2 weighed sequences (CISS, DRIVE……)
MECKEL CAVE PLASTIC EXTENSION ALONG 5th CRANIAL NERVE





CISS





AJNR Am J Neuroradiol 42:961–68 May 2021
Courtesy of Prof G Morana

Criticità

- Sede della lesione
- Radicalità dell'intervento

- Protocolli RM post-operatori adeguati per valutare l'eventuale residuo (sede, strutture adiacenti e dimensioni)
- Importanza dell'uso delle sequenze volumetriche T2 pesate (CISS, DRIVE……)
- Indicazioni di follow-up in caso dubbio

Ependimoma Stratum II Spagna

In corso, sarà conclusa entro fine aprile

Aperto braccio osservazionale e studio gemello BIOMECA Italy – tumor tissue collected

- Febbraio 2026:
- 107 FFPE of which 79 molecularly classified;

- Additional 65: unstained slides/rolls - limited material

SIOPE DIPG/DMG Registry Update

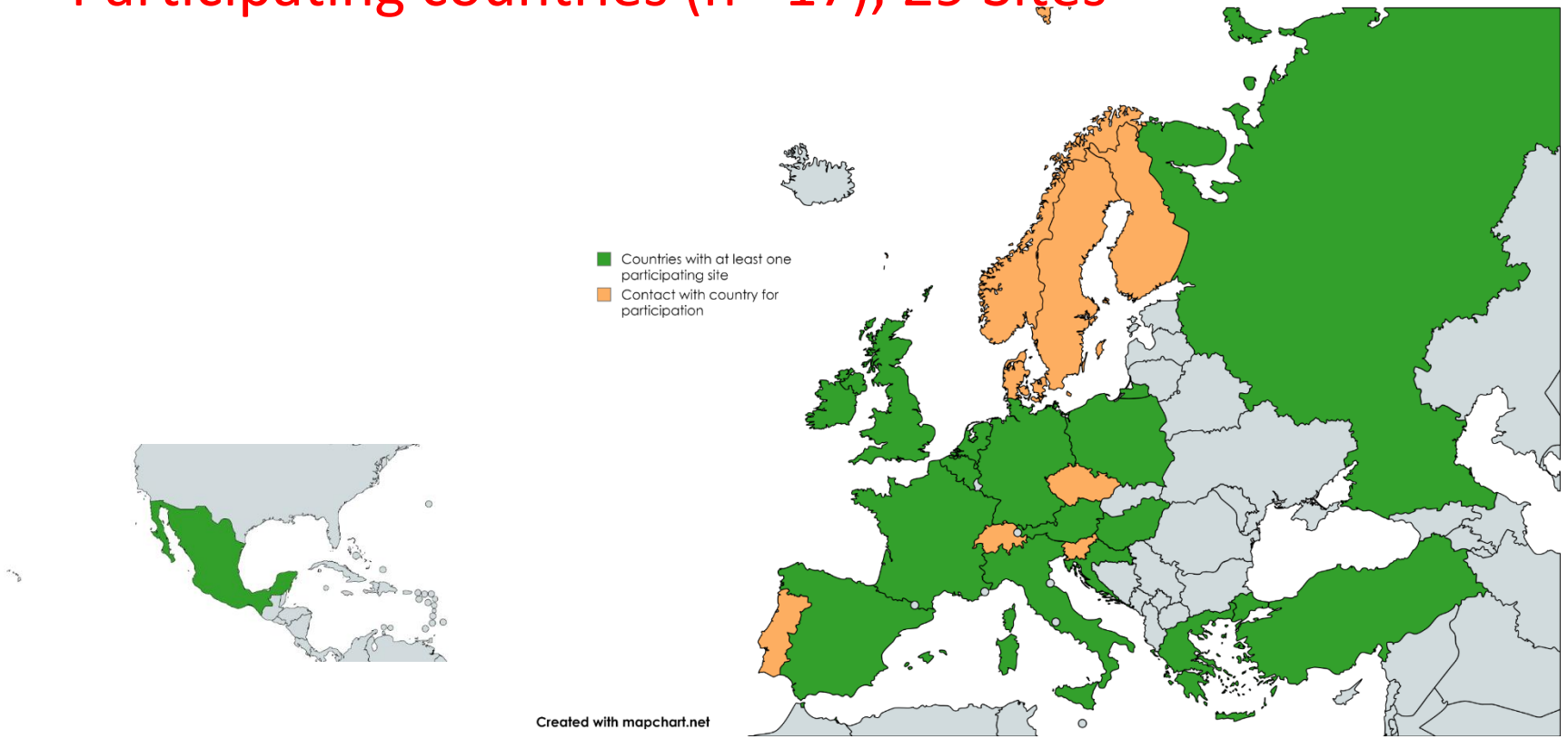
Ongoing efforts of building a European database for collaborative research initiatives, aimed at improving the care for DIPG and DMG patients

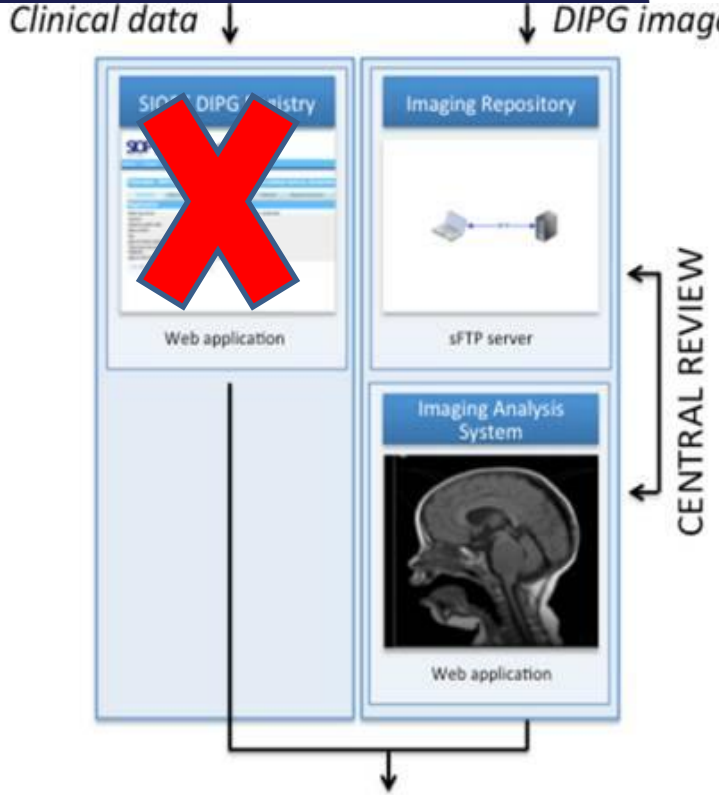
all of us can contribute to this!

GdL AIEOP Tumori Cerebrali 25/02/2026

SIOP Europe
the European Society for Paediatric Oncology

Participating countries (n= 17), 29 Sites





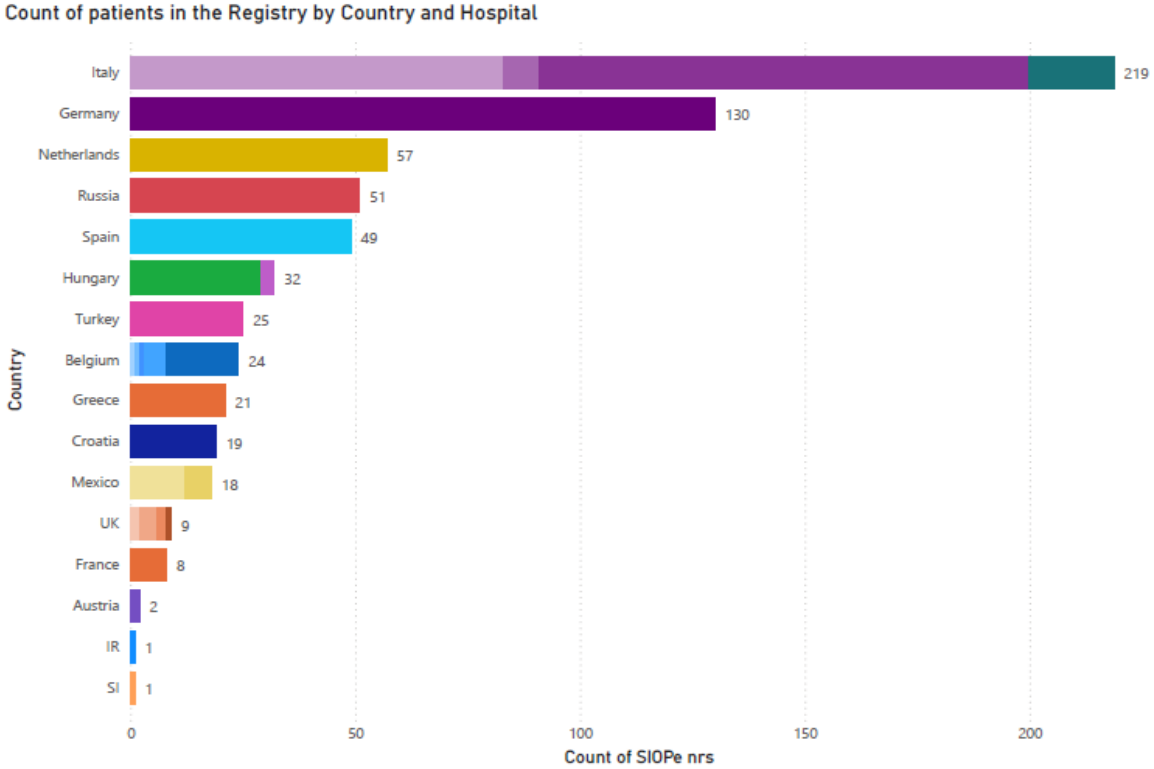
DIPG/DMG COLLABORATIVE

Princess máxima center pediatric oncology

Exhaustivity check and quality control of the data*

↓

Data storage within the SIOPE DIPG Registry database



Patients registered in ALEA

666

Patients with diagnostic imaging registered in MDPE

276

Countries including pts

17

Sites including patients

29

Hospital	Count of Patients registered in ALEA	Count of MDPE registered=Y	Count of Diagnostic imaging=Y
Agia Sophia Childen's Hospital	21	6	5
CHI, Crumlin	1		
Children's Hospital Zagreb	19	19	19
CHR Citadelle	1		
CHU Strasbourg	8		
Clinique Universitaire Saint Luc, Bussels	1		
Fondazione IRCCS Istituto dei Tumori di Milano	83	82	81
GPOH	130		
Hospital de Pediatría del Centro Médico Nacional Siglo XXI	12		
Instituto Nacional de Pediatria	6		
Istanbul University, Oncology Institute	25	4	4
Leeds Children's Hospital NHS Foundation Trust	2	1	1
Liverpool Alder Hey Children's Hospital	4		
Medical University of Graz	2		
Meyer Children's Hospital, Florence	8		
Ospedale Pediatrico Bambino Gesù, Rome	109	83	82
Prinses Máxima Centrum	57	48	47
Russian Scientific Center of Roentgenradiology	51	25	18
RVI:Great North Children's Hospital, Newcastle	2	1	1
Santobono-Pausilipon Childrens Hospital, Naples	19	2	1
Total	**666**	**292**	**276**

ITALIAN PATIENTS

219/666 33%

Total enrollment: n = 1179
Retrospective (n = 634) + Prospective (n = 545)

VOLUME 36 · NUMBER 19 · JULY 1, 2018
JOURNAL OF CLINICAL ONCOLOGY
ORIGINAL REPORT

Clinical, Radiologic, Pathologic, and Molecular Characteristics of Long-Term Survivors of Diffuse Intrinsic Pontine Glioma (DIPG): A Collaborative Report From the International and European Society for Pediatric Oncology DIPG Registries

Lindsey M. Hoffman, S
Marion Hoffmann, Ad
Goldman, Sarah Leary,
Kieran, Jane Minturn,
Chintagumpala, Anne S
Hetal Dholaria, Renee
Boddaert, Pascale Varle
Piergiorgio Modena, M
Dominik Sturm, Stefan
Hargrave, Guirish A. S
Simon Bailey, Veronica
Warmuth-Metz, James

MATERIALS AND METHODS

Study Population

The study was approved by the institutional review board at Cincinnati Children's Hospital Medical Center and included 1,130 patients with radiographically confirmed DIPG diagnosed from 1990 to 2015. IDIPGR patients (n = 409) were age 0 to 27 years from the United States, Canada, and Australia. SIOPE-DIPGR patients (n = 721) were age 0 to 21 years from the Netherlands, Germany, Austria, Switzerland, Italy, France, the United Kingdom, and Croatia. Patients were referred to the registries as previously described.[6,7] Exclusion criteria are listed in Figure 1. No patients with neurofibromatosis type 1 were included.

Table 1. Retrospective Registry Patients

Country	#patients
Germany	278
Netherlands	114
France	113
Italy	79
UK	43
Croatia	7
TOTAL	**634**

BRAIN TUMOUR GROUP

SIOPE-HGG-01

SIOPE-BTG-HGG-01

International cooperative randomized trial of the SIOPE HGG/DIPG Working Group for the treatment of newly diagnosed and recurrent high-grade gliomas in children, adolescents, and young adults

Responsible Pediatric oncologists: Veronica Biassoni, Michael Karremann, Christof Kramm, and Maura Massimino

SIOPE HGG/DIPG Working Group
GESELLSCHAFT FÜR PÄDIATRISCHE ONKOLOGIE UND HÄMATOLOGIE GPOH
FONDAZIONE IRCCS ISTITUTO NAZIONALE DEI TUMORI
UNIVERSITÄTSMEDIZIN GÖTTINGEN UMG
UMM UNIVERSITÄTSMEDIZIN MANNHEIM

General aims of the trial

Option to participate in a clinical trial **close to home**

Establishing a **European platform** with high quality standards

Targeting the **microenvironment**

Study alternative **radiotherapy** concepts

Overview

Patient with High Grade Glioma
Diffuse HGG, non-diffuse HGG, DIPG and Gliomatosis cerebri

y diagnosis and age 3 to 21 years		Relapse/Progression and age 3 to 24 years	
Screening: central review process (2-3 weeks) (neuropathology and/or neuroradiology)		**Screening**: central review process (2-3 weeks) (neuroradiology ± neuropathology)	
DIPG or **DMG**, **non-metastatic**	**Other HGG** incl. metastatic DMG/DIPG	**DIPG** or **DMG**	**Other HGG** non-DMG/non-DIPG
⇩	⇩	⇩	⇩
Stratum 1	**Stratum 2**	**Stratum 3**	**Stratum 4**
⇩	⇩	⇩	⇩
Randomization: CSI vs. focal radiotherapy 158 patients Estimated recruitment 4.08 years	**Randomization:** Perampanel vs. Metformin 141 patients Estimated recruitment 3,85 years	**Randomization:** Nimo/Vino vs. Vino 129 patients Estimated recruitment 3.30 years	**Observational trial:** Re-irradiation + mebendazole 100 patients Estimated recruitment 4.00 years
Treatment: 13 months	**Treatment:** 13 months	**Treatment:** 12 months	**Treatment:** 12 months
Minimum follow up: 24 months from time of diagnosis		**Minimum follow up:** 24 months from time of preceeding progression	

Formal steps regarding side projects

Summer 26

Assessment by authorities and IRBs during CTIS process

Collection of scientific samples is objective of the trial. Abstracts of the side projects as well as patient information and informed consent form for data collection for the specific projects will be evaluated.

(IRB) approval for the trial applies also for the side projects

End of 26/early 27

Inclusion of first patient

Once the trial is approved, samples can be collected from every patient after IC for the specific project.

After approval

Request funding for side projects and start analyses

There is no time until side projects need to start analyses or need to be finalized. Funding for condution of side projects by investigators

Countries involved

Austria
Germany
Italy
Switzerland
The Netherlands
Spain

Recruiting: 4 years

Happy to initiate the trial in further countries, once it has been approved by the authorities

LOGGIC Core
LOGGIC: LOw Grade Glioma In Children

LOGGIC CORE

Bio Clinical Data Bank

SIOPe LGG Working Group

LOGGIC Core

LOGGIC: LOw Grade Glioma In Children

Coordinatore Internazionale

Germania

- Hopp Children's Cancer Center at the NCT Heidelberg (KiTZ)

- German Cancer Research Center (DKFZ)

Coordinamento Nazionale

Italia

- Rappresentante del Promotore: AIEOP
- Centro Coordinatore: Ospedale Pediatrico Bambino Gesù

LOGGIC Core
LOGGIC: LOw Grade Glioma In Children

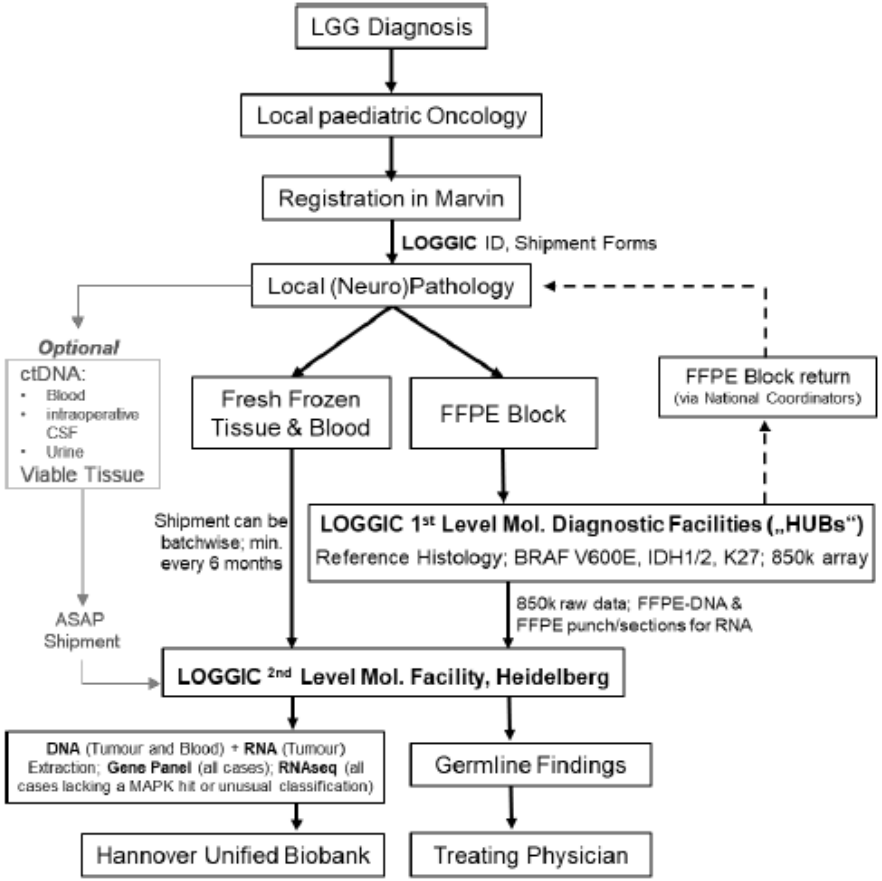
Obiettivi dello studio

1) To provide **tumour characteristics of LGG patients** for a potential inclusion into trials like LOGGIC Europe Trial by collecting tumour material and baseline molecular, histological, and clinical data.
2) To **improve diagnostic accuracy for all LGG patients** included in LOGGIC Core by adding molecular information to reference histologic/radiologic evaluation.
3) To **identify clinically relevant targets** for possible future targeted treatments in clinical trials.
4) To **identify tumour tissue derived biomarkers** for treatment response and natural course of disease by linking molecular with radiological and clinical follow-up data.
5) To **perform liquid biopsy** analysis on different sources including blood, intraoperative CSF and urine as a biomarker for diagnosis and response to treatment as well as other exploratory biological studies.
6) To set up an **ex-vivo drug testing** and model development pipeline on viable tumour tissue.
7) To establish a LOGGIC biobank and use the Hannover Unified Biobank to store extracted DNA/RNA.

1th level molecular facility

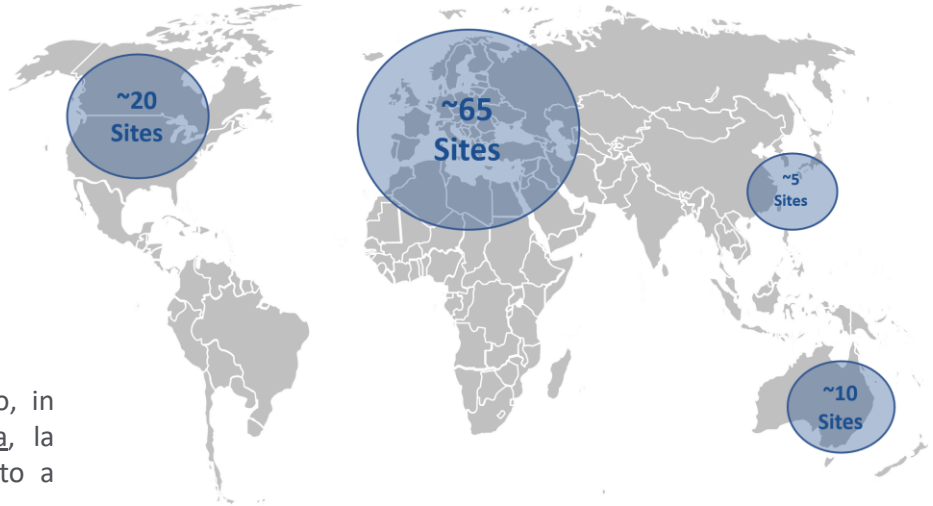
- La centralizzazione sarà Sapienza
- I centri invieranno come da pratica clinica tutto il materiale richiesto in Sapienza
- Primo livello: invio solo della paraffina
- Alla conferma diagnostica Sapienza invia a OPBG per diagnostica molecolare
- Alla conferma di arruolabilità i centri inviano a Sapienza il resto del materiale congelato
- Lo studio è sponsorizzato da FUV e con questi fondi AIEOP rimborserà trasporti e analisi

Shipment: da centro a Sapienza e da Sapienza ad Heidelberg



Collaborazione tra Day One e il consorzio LOGGIC

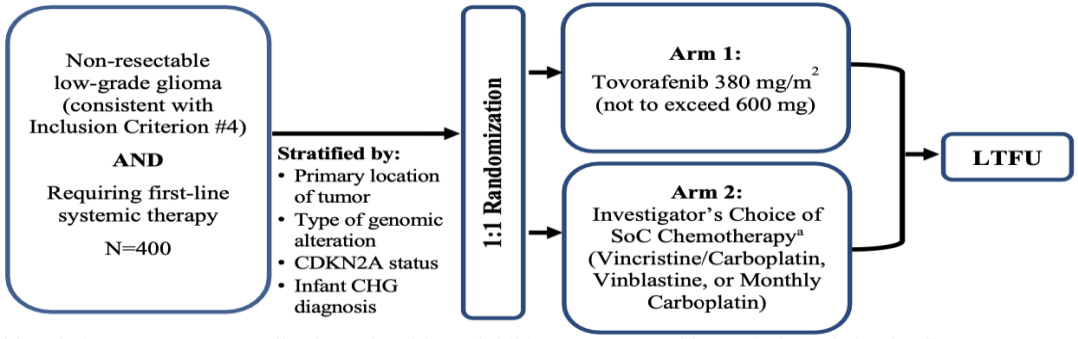
- Partecipazione di **circa 100 istituti**
- **Periodo di arruolamento** – 3 anni; durata dello studio - 7 anni
- **Aspettative di arruolamento**:
  - 2 pazienti per centro/anno



- **Disegno dello studio:** studio di fase 3, a 2 bracci, randomizzato, in aperto, multicentrico, internazionale, volta a valutare l'efficacia, la sicurezza e la tollerabilità di Tovorafenib in monoterapia rispetto a chemioterapia SoC
- **Dimensione prevista del campione:** N=400 pazienti naïve al trattamento, di età <25 anni con pLGG, con un'alterazione attivante di RAF

KiTZ Hopp Children's Cancer Center Heidelberg | German Cancer Research Center (DKFZ) Heidelberg University Hospital Heidelberg University

LOGGIC EUROPE
LOGGIC: LOw Grade Glioma In Children



Abbreviations: CDKN2A, cyclin-dependent kinase inhibitor 2A; CHG, chiasmatic-hypothalamic glioma; COG-V/C, Children's Oncology Group – Vincristine/Carboplatin; LTFU, long-term follow-up; SIOPe-LGG-V/C, International Society for Paediatric Oncology – Low-Grade Glioma Vincristine/Carboplatin; SoC, Standard of Care.
[a] Arm 2: COG–V/C, SIOPe-LGG–V/C, vinblastine, or monthly carboplatin (see Section 5).

- **Trattamento:** Tovorafenib (Braccio 1) o chemioterapia SoC (Braccio 2) con vincristina/carboplatino (COG, SIOPe), vinblastina, o carboplatino mensile
- I pazienti che progrediscono nel braccio della chemioterapia SoC durante o dopo il trattamento possono effettuare il cross-over per ricevere Tovorafenib
- Lo studio prevede:
  - Fase di screening
  - Fase di trattamento
  - Visita di fine trattamento
  - Visita di follow-up a 30 giorni
  - Periodo di follow-up a lungo termine

Obiettivo primario:

- Confrontare il **tasso di risposta obiettiva (ORR) secondo i criteri della Valutazione della risposta in neuro-oncologia per i gliomi di basso grado (RANO-LGG)** valutato dal comitato di revisione indipendente (IRC) di DAY101 in monoterapia rispetto alla chemioterapia standard di cura (SoC), in pazienti con glioma pediatrico di basso grado con un'alterazione RAF che richiede una terapia sistemica di prima linea

Obiettivi secondari principali:

- Confrontare la sopravvivenza libera da progressione (PFS), in base alla valutazione dell'IRC, di DAY101 in monoterapia rispetto alla chemioterapia SoC secondo i criteri RANO-LGG.
- Confrontare la sopravvivenza libera da eventi (EFS), in base alla valutazione dell'IRC, di Tovorafenib in monoterapia rispetto alla chemioterapia SoC secondo i criteri RAPNO.
- Confrontare la sopravvivenza complessiva (OS) di DAY101 in monoterapia rispetto alla chemioterapia SoC.

Obiettivi secondari dello studio

Sicurezza ed esiti funzionali

- Confrontare la sicurezza e la tollerabilità della monoterapia con Tovorafenib rispetto alla chemioterapia SoC.
- Confrontare le variazioni nella funzione neurologica e nel funzionamento adattivo tra Tovorafenib e chemioterapia SoC utilizzando *Vineland Adaptive Behavior Scale* (VABS).
- Confrontare le variazioni negli esiti della funzione visive della monoterapia con Tovorafenib e chemioterapia SoC in pazienti con glioma delle vie ottiche (OPG).

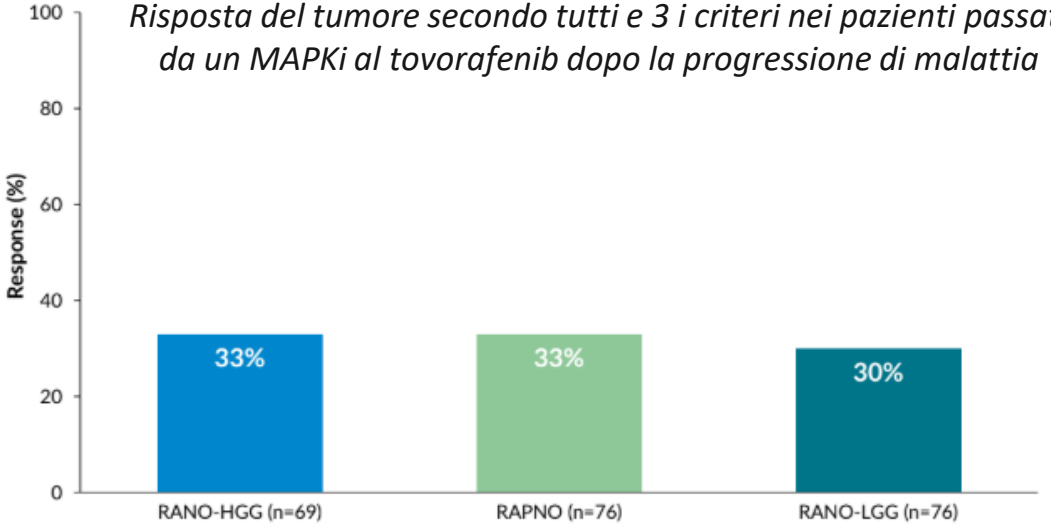
Qualità di vita

- Valutare la qualità della vita correlate allo stato di salute (HRQoL) con monoterapia Tovorafenib rispetto a chemioterapia SoC utilizzando la batteria di test Patient-Reported Outcomes Measurement Information System (PROMIS®)

Nello studio FIREFLY – 1

Analisi post hoc: Risposte tumorali al Tovorafenib in pazienti progrediti con MAPKi come terapia precedente più recente

Risposta del tumore secondo tutti e 3 i criteri nei pazienti passati da un MAPKi al tovorafenib dopo la progressione di malattia



Tossicità riscontrate

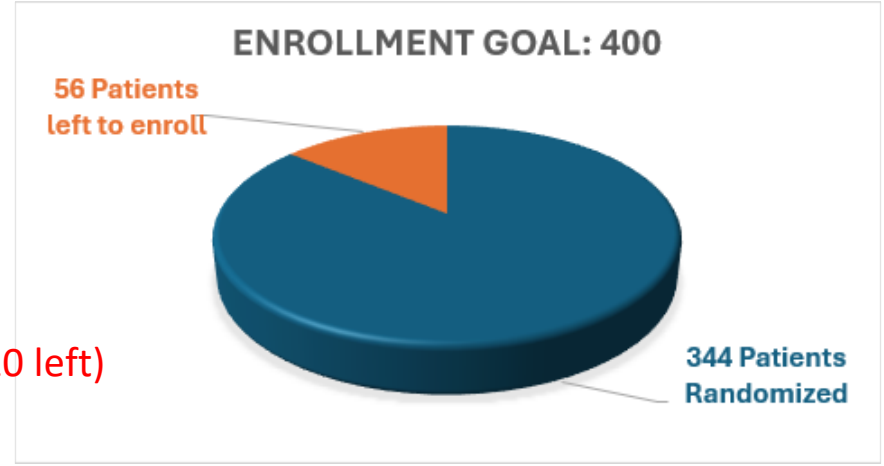
Tossicità da Chemioterapia SoC	Tossicità da Tovorafenib
• Anemia • Necessità di trasfusioni di piastrine	• Tossicità cutanea • Arresto della crescita • **Sanguinamento di una cisti subaracnoidea** a distanza di 5 mesi dall'avvio del trattamento (Tovorafenib). L'evento ha richiesto un intervento NCH, risolto senza esiti

Stato dell'arruolamento per centro

Centri arruolati in Italia

CENTRO	TOTALE	TRATTATI con CHEMIOTERAPIA SoC	TRATTATI con TOVORAFENIB	CROSS-OVER (Chemioterapia → Tovorafenib)
Torino	3	2	1	0
Milano	7	6	**1**	**(1)**
Genova	7	5	2	0
Padova	**(1)**	0	0	0
Udine	3 + **(1)**	3	0	**1**
Roma	10	6	4	**2**
Napoli	1	1	0	0
Bari	3	3	0	0
Totale - Italia	**30**	**22**	**8**	**2/3**

Stato dell'arruolamento: 344 pazienti randomizzati su 400 (dati aggiornati al 31 gennaio 2025)



(April: only 10 left)

NF1-LGG BTG group
Breve Aggiornamento

- **Chiusura in US studio COG ACNS 1381 (selumetinib *Vs* Vcr-Carbo) per arruolamento non adeguato (75pz inclusi in US)**
  - Facilità accesso farmaci off-trial in US
  - Mai aperto per difficoltà burocratiche insormontabili in Europa/UK

- **Unica alternativa possibile trial con MEKi: Mirdametinib ?**
  - Dati studio fase I/II SJ901 in LGG (NF1+/-)
  - Discussioni preliminari positive con Springworks

- **Vincristina/carbo (SIOP-LGG) rimane lo *standard of care***
  - Uso **vinblastina** *single agent* sempre più accettato per migliore profilo tollerabilità
  - Ruolo del **Bevacizumab** in casi selezionati* (648?)

* Es calo del visus/campo visivo

EPILOGUE: Phase I/II Combination Umbrella Trial in Relapsed Pediatric Low Grade Glioma

Gruppo di Lavoro AIEOP
Tumori Cerebrali

25 febbraio 2026

Elisabetta Schiavello

EPILOGUE: Phase I/II Combination Umbrella Trial in Relapsed Pediatric Low Grade Glioma (pLGG)

KiTZ Hopp Children's Cancer Center Heidelberg

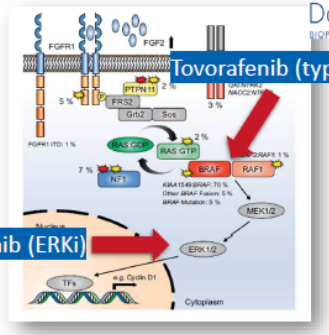
- **Rationale:** pLGG — a MAPK-driven disease. Combining targeted and conventional therapy may improve durability

Olszanski AJ et al. Annals of Oncol 2017
Sun Y et al. Neuro Oncol 2017
van Tilburg C et al. Neuro Oncol 2022
Kilburn LB et al. Nat Med 2023

Day One BIOPHARMACEUTICALS

Jones et al. Cancer Res 2008
Pfister et al. JCI 2008
Jones et al. Nature Genetics 2013
Zhang et al. Nature Genetic 2013
Reitman et al. Nature Comm 2019
Ryall et al. Cancer Cell 2020
Usta et al. Mol Cancer Ther 2020
Sigaud et al. Neuro Oncol 2022
Sigaud et al. Nature Comm 2023

BIOMED VALLEY DISCOVERIES



Tovorafenib (type II RAFi)

Ulixertinib (ERKi)

Oncogene-induced senescence (OIS)

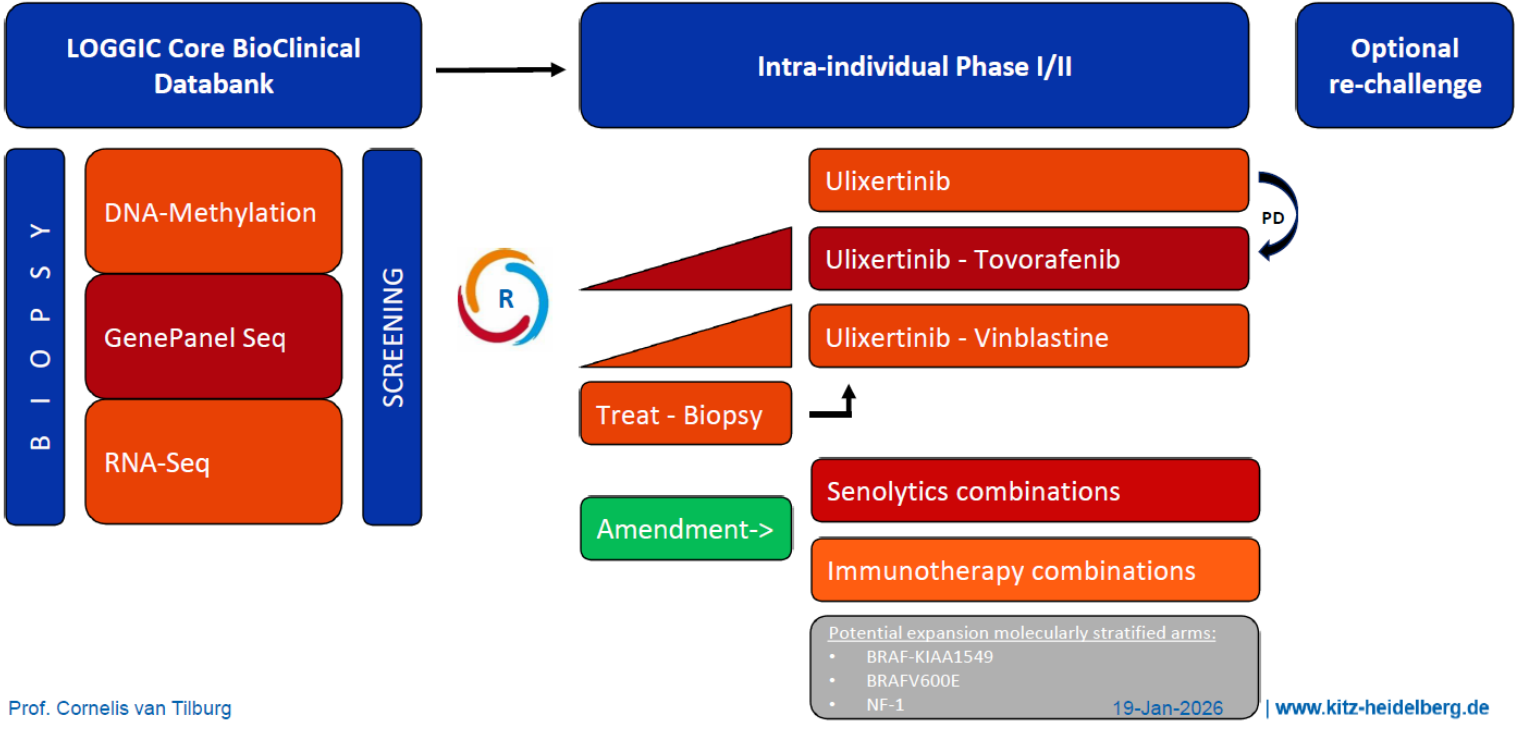
Bcl-Xli

Jacob et al. Clin Cancer Res 2011
Buhl et al. Clin Cancer Res 2018
Reitman et al. Nature Comm 2019
Selt et al., Neuro Oncol 2022

ITCC
INNOVATIVE THERAPIES FOR CHILDREN AND ADOLESCENTS WITH CANCER

EPILOGUE Trial Design

KiTZ Hopp Children's Cancer Center Heidelberg



Timelines and Funding

Trial Duration and Dates (first 3 arms)

- **CTIS submission:19 Nov 2025**
- Recruitment duration: 2 years
- Treatment duration: max. 48 weeks
- Minimal follow-up duration after EOT of first treatment course: 1 year
- Total trial duration: 4 years
- Trial initiation & first patient in: April 2026
- CTIS second submission: Q3 2026
- Second trial initiation & first patient in: Q4 2026

Sites and Countries

- 22 participating sites across the ITCC network
- **First CTIS submission:** Austria (1), Czech Republic (1), Denmark (1), Germany (5), Sweden (1)
- **National submissions after first CTIS approval:** Australia (5), United Kingdom (4)
- Second CTIS submission: The Netherlands (1), Spain (2)
- Italy and Poland (pending ongoing discussions)

Drug Support and Financing

- BioMed Valley Discoveries (ulixertinib)
- Day One Biopharmaceuticals (tovorafenib)
- Everest startup funding from the Brain Tumor Charity
- Dietmar Hopp Foundation

SIOPe ATRT01

An international prospective umbrella trial for children with atypical teratoid/rhabdoid tumours (ATRT) including **A randomized phase III study evaluating the non-inferiority of three courses of high-dose chemotherapy (HDCT) compared to focal radiotherapy as consolidation therapy**

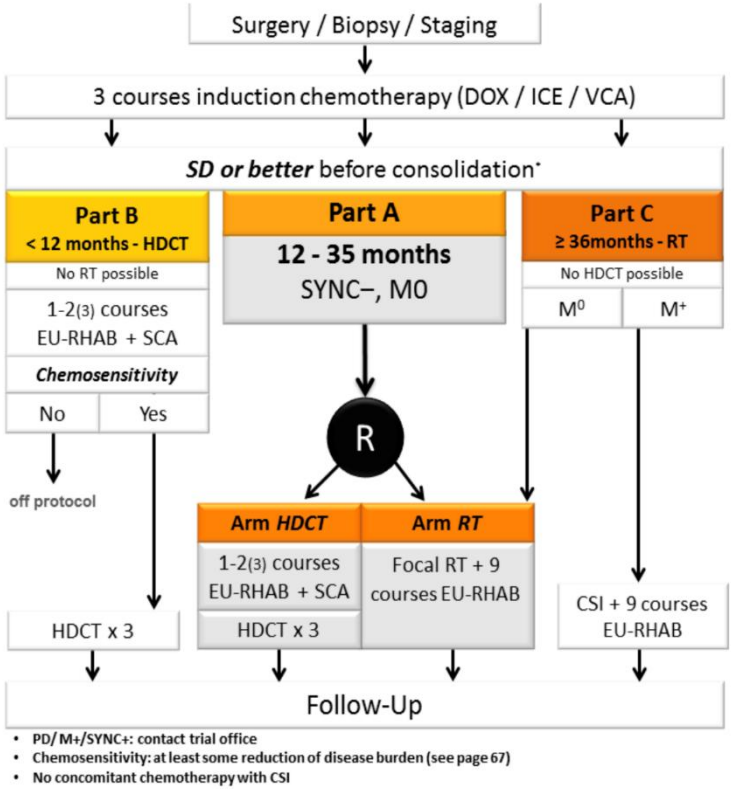
Protocol update

Protocollo SIOPE ATRT01 - timeline

Centro:	Dipartimento	Città	Principal investigator	Stato di attivazione
IRCCS Ospedale Pediatrico "Bambino Gesù"	Area Studi Clinici Oncoematologici e Terapie Cellulari	Roma	Angela Mastronuzzi	Attivo (NCC e centro)
IRCCS Azienda Ospedaliero-Universitaria di Bologna, Policlinico Sant'Orsola	Oncologia ed Ematologia Pediatrica "Lalla Seràgnoli" Clinica Pediatrica	Bologna	Fraia Melchionda	Negoziazione contratto in corso
A.O.U. Città della Salute e della Scienza di Torino - Presidio Infantile Regina Margherita	SC Oncoematologica Pediatrica e Centro Trapianti	Torino	Franca Fagioli	Negoziazione contratto in corso
AORN Santobono-Pausilipon	Oncoematologia Pediatrica	Napoli	Lucia Quaglietta	Attivo
IRCCS Istituto Nazionale dei Tumori di Milano	S.C. Pediatria Oncologica	Milano	Elisabetta Schiavello	Attivo
Azienda Ospedale Università Padova	UOC Oncoematologia Pediatrica	Padova	Elisabetta Viscardi	SIV programmata

Flowchart protocollo

- Umbrella Trial
  - Registrazione
  - Chemioterapia di Induzione
- SIOPE ATRT01
  - Registrazione
  - Stratificazione
  - *Randomizzazione (solo Group A)*
  - Consolidamento
  - Follow-up



Centralizzazioni

Tessuto tumorale FFPE (+ congelato)	Università La Sapienza (dr.ssa Manila Antonelli)	Diagnosi (**entro Fine Induzione** – NON vincolante per arruolamento in Trial Umbrella!) *In caso di re-intervento*
Liquor	Università La Sapienza (dr.ssa Manila Antonelli)	Diagnosi **Induzione**
Neuroimaging	OPBG (dr.ssa G. Stefania Colafati)	Diagnosi **Fine Induzione** *Post re-intervento* Durante consolidamento (in base a stratificazione)
Neurochirurgia (second opinion)	OPBG (dr. Andrea Carai)	Possibilità di re-intervento (Post-diagnosi? **Dopo Induzione?**)
Radioterapia (planning)	INT (dr.ssa Sabina Vennarini)	Consolidamento (solo per Group A – random Arm RT)

Indispensabili per arruolamento al trial ATRT01 (dopo Induzione)

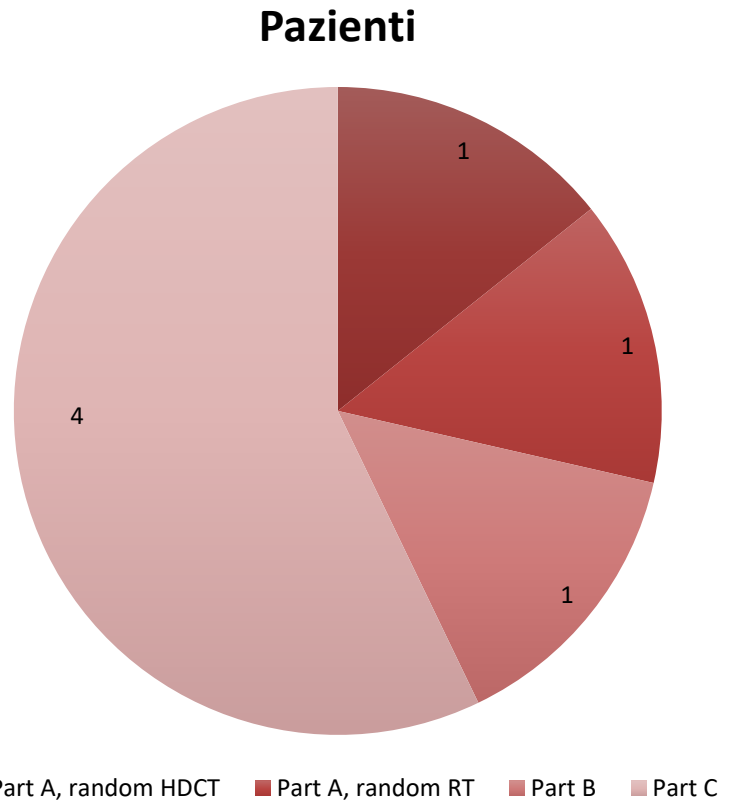
SIOPE ATRT - OPBG Center

10 pazienti arruolati:

- 1 paziente incluso solo nell'Umbrella Trial (nessun trattamento attivo a causa di gravissime condizioni neurologiche)
- 1 drop out per diagnosi

<u>Totale pazienti valutabili: 8</u>

→ 1 deceduta per PD dopo induzione
→ 4 hanno completato il trattamento
→ 3 pazienti in corso



Bambino Gesù
OSPEDALE PEDIATRICO

Phase I study of anti-GD2 Chimeric Antigen Receptor-Expressing T cells in pediatric and young adult patients affected by relapsed/refractory central nervous system Tumors

P.I. Prof Franco Locatelli

ClinicalTrials.gov Identifier: NCT05298995

Clinical Trial Design

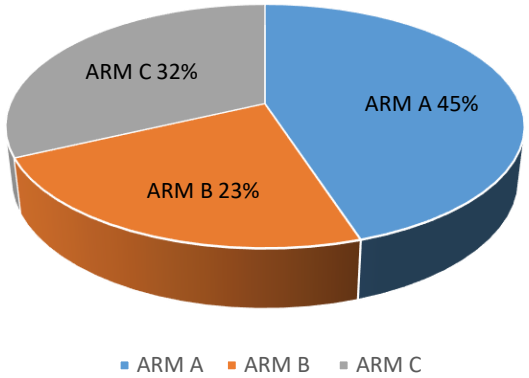
- Tre braccia di trattamento:
  - **ARM A: MB/altri tumori embrionari**
  - **ARM B: HGG emisferici**
  - **ARM C: HGG talamici, DMG e altri rari tumori del SNC non inclusi nell'arm A e B**

- Dose levels per ogni braccio:
  - DL1: 0.25 x 10^6 cells/kg di CAR+ T cells
  - DL2: 0.5 x 10^6 cells/kg di CAR+ T cells
  - **DL3: 1.0 x 10^6 cells/kg di CAR+ T cells**
  - DL4: 3.0 x 10^6 cells/kg di CAR+ T cells
  - DL5: 6.0 x 10^6 cells/kg di CAR+ T cells

- Arruolamento parallelo in sequenza per ciascun braccio: una volta concluso l'arruolamento della coorte di pazienti DL3 del braccio A, in assenza di DLT, sarà attivato un arruolamento parallelo come segue: coorte DL4 di pazienti dell'ARM A e coorte DL3 di pazienti dell'ARM B. La stessa strategia sarà applicata all'ARM C.
- **Si prevede di includere 27 pazienti**, secondo lo schema di escalation della dose 3+3, fino a un massimo di **54 pazienti** (18 pazienti per braccio) arruolati in un periodo di 24 mesi.

Aggiornamento arruolamento pazienti

- *Data apertura protocollo: 09/11/2023*
- *Arruolamento attuale: 22 pazienti*
- *Pazienti trattati attualmente: 17 pazienti*



AIEOP
ASSOCIAZIONE ITALIANA EMATOLOGIA ONCOLOGIA PEDIATRICA

GdL – Tumori del Sistema Nervoso Centrale (SNC)

Studio spontaneo osservazionale retrospettivo sui Tumori del Sistema Nervoso Centrale e Studio spontaneo osservazionale prospettico sui Tumori del Sistema Nervoso Centrale

Titolo breve:
DI-TSNC
(Dati Italiani tumori del Sistema Nervoso Centrale)

Associazione Italiana Ematologia Oncologica Pediatrica

Codice Protocollo: DI-TSNC

35 Centri candidati

- 25 Centri «attivati»
  - 10 retrospettivo + prospettico
  - 12 retrospettivo con CI + prospettico
  - 3 in corso di definizione modalità di partecipazione

AIEOP
ASSOCIAZIONE ITALIANA EMATOLOGIA ONCOLOGIA PEDIATRICA

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Studio spontaneo osservazionale retrospettivo sui
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Titolo breve:
DI-TSNC
(Dati Italiani tumori del Sistema Nervoso Centrale)

Associazione Italiana Ematologia Oncologica Pediatrica

Codice Protocollo: DI-TSNC

Criticità

- Passaggio CINECA → DATA RIVER
  - Emendamento al Protocollo
  - Revisione dei consensi
- Visibilità delle schede di FU

In fieri

- Analisi preliminare dei dati finora raccolti

ALTRI PROGETTI

studio retrospettivo osservazionale OPG (Lucia Quaglietta)
a. Saranno raccolti i dati endocrinologici, visivi e radiologici dei pazienti affetti da OPG (3 gruppi – OPG NF1 relati trattati/non obiettivo principale è valutare la PFS visiva ed endocrinologica.
c. dati di OCT del n. ottico (per le valutazioni visive) e della qualità della vita.

craniofaringioma

Proposta Gruppo multidisciplinare Gaslini
-neuroncologi ed endocrinologi:

- Avviare prima registro Italiano
- Ispirato a quello tedesco
- Unirsi in un secondo momento al Tedesco/Europeo

Discussione nazionale di casi di Neuroncologia pediatrica

- Scopo: confronto/conforto non vincolante su casi complessi
- Kick-off 28 ottobre 2025
- 3 incontri
- 11 casi discussi
- Casi presentati da 8 Centri italiani

Prof.ssa Natascia Di Iorgi
endocrinologa
natasciadiiorgi@gaslini.org

Dott. Antonio Verrico
neuroncologo
antonioverrico@gaslini.org

& linee guida /12/4/2026
Gruppo cooperativo da SINCH

Revisione radioterapica: ricadute locali ependimoma nel protocollo AIEOP 2

Progetto Dr.ssa M.Massimino-Dr.M.Mascarin –Dr.ssa S.Vennarini

Riunione Gruppo di Lavoro AIEOP Tumori Cerebrali – 25/02/2026

Dr.ssa Sabina Vennarini
S.S. Radioterapia Pediatrica
Fondazione IRCCS Istituto Nazionale Tumori
sabina.vennarini@istitutotumori.mi.it

FONDAZIONE IRCCS ISTITUTO NAZIONALE DEI TUMORI

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Dove recidivano gli Ependimomi?

- Studio Retrospettivo Multicentrico Nazionale (aperto a chi vuole aderire)

- Pazienti R0-AIEOP 2; *localizzazione della 1 recidiva in quale isodose di radioterapia*

- Casistica Raccolta:

- Database INT: 14 Pt/9 Pt in cui è stato possibile recuperare il piano di trattamento

- Database Genova : 1 Pt

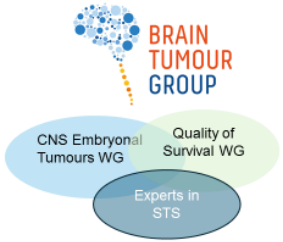
- Database Roma: 1 Pt (non ancora inserito in attesa di documentazione completa)

Ototossicità
Gruppo di Lavoro SIOPE - BTG

Chair: Rebecca Hill (UK)

Components

- Johnathan Abrams, Antony Penn (UK)
- Enrico Opocher, Elisabetta Schiavello (Italy)
- Astrid Sehested (Denmark)
- Katrin Scheinemann (Switzerland, QOS/Pancare member)
- Niki Roka (Greece)
- Magnus Sabel, Aron Onerup (Sweden)



Gruppo di lavoro ototossicità

AIEOP
ASSOCIAZIONE ITALIANA ONATOLOGIA ONCOLOGIA PEDIATRICA

2° Incontro del Gruppo di Lavoro AIEOP Ototossicità
22 Gennaio 2026
Orario 10.45 – 16.00

Agenda Definitiva

- 10.45 – 11.00 Benvenuto, Introduzione ed Obiettivi (Enrica/Michela)
- 11.00 - 11.20 Fattori di Rischio per ototossicità (Antonio/Alberto)
- 11.15 – 13.15 discussione del rischio di ototossicità e fattibilità strategie di prevenzione/mitigazione nelle diverse patologie (tutti)
  - Sarcomi ossei
  - Tumori a cellule germinali
  - Tumori non Epatoblastoma
  - Neuroblastoma
  - Medulloblastoma
  - Tumori a cell germinali intracranici
- 13.15 – 14.00 pranzo
- 14.00 Inizio condivisione documento di raccomandazioni su ototossicità e strategie di prevenzione/mitigazione – 60'
  - schede di patologia (rischio/beneficio)
  - modalità prescrizione, infusione e terapia di supporto in corso di STS (Virginia/Alberto)
- 15.00 – Aggiornamento su progetti e proposte, prossimi appuntamenti
  - Raccolta dati retrospettivi (F. Calò, C. Novara Ts, Romano)
  - Proposte per un monitoraggio a lungo termine – Ruggiero/Romano/Bivetto
  - QOL survey (Pioneli/Bivetto)
  - Prossimi incontro alle (pre-)Giornate tumori solidi: PD 11/3/26, poi AIEOP Lecce?
- 16.00 Fine incontro

per richiesta spese viaggio scrivere a segreteria@aieop.org

Proposte al GDL-SNC su ototox

- **Feedback/aggiornamento** sul lavoro nel gruppo SIOP-BTG
- **Survey** su frequenza, monitoraggio e gestione ototossicità nei paz con tumori SNC e cisplatino (Medulloblastoma, NGGCT, LGG?)
- **Incontro multidisciplinare** (audiologi, RT, oncologi) dedicato ad ototox nei tumori **SNC ?**

Produzione scientifica

30 lavori nel 2025