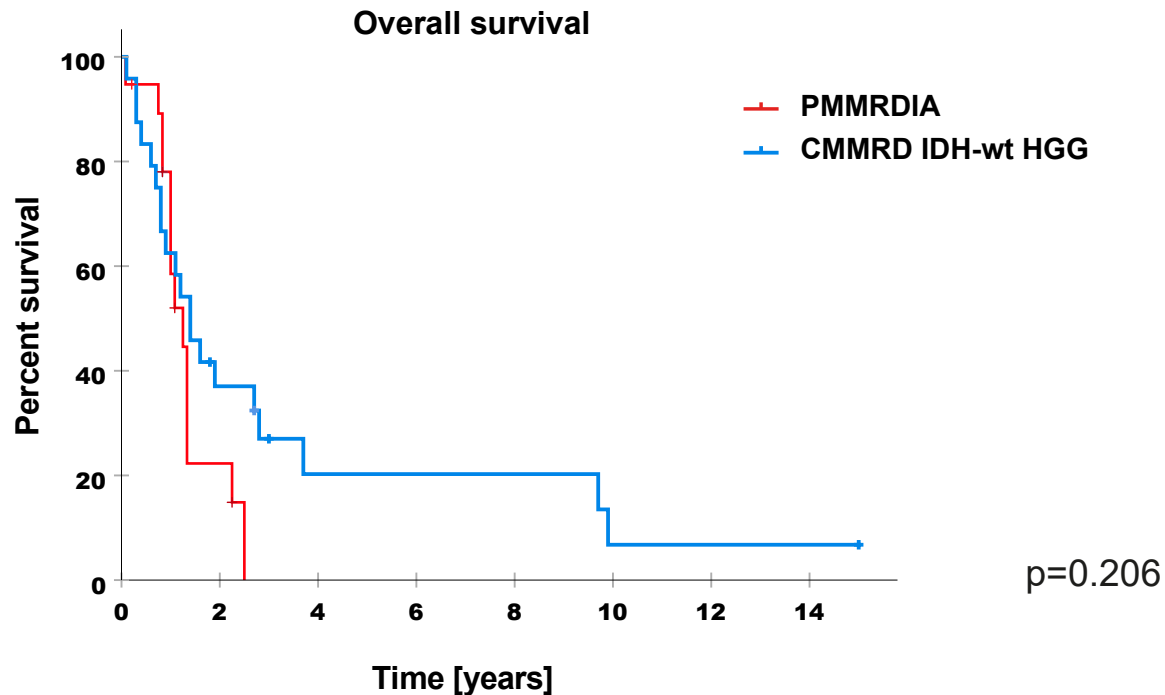


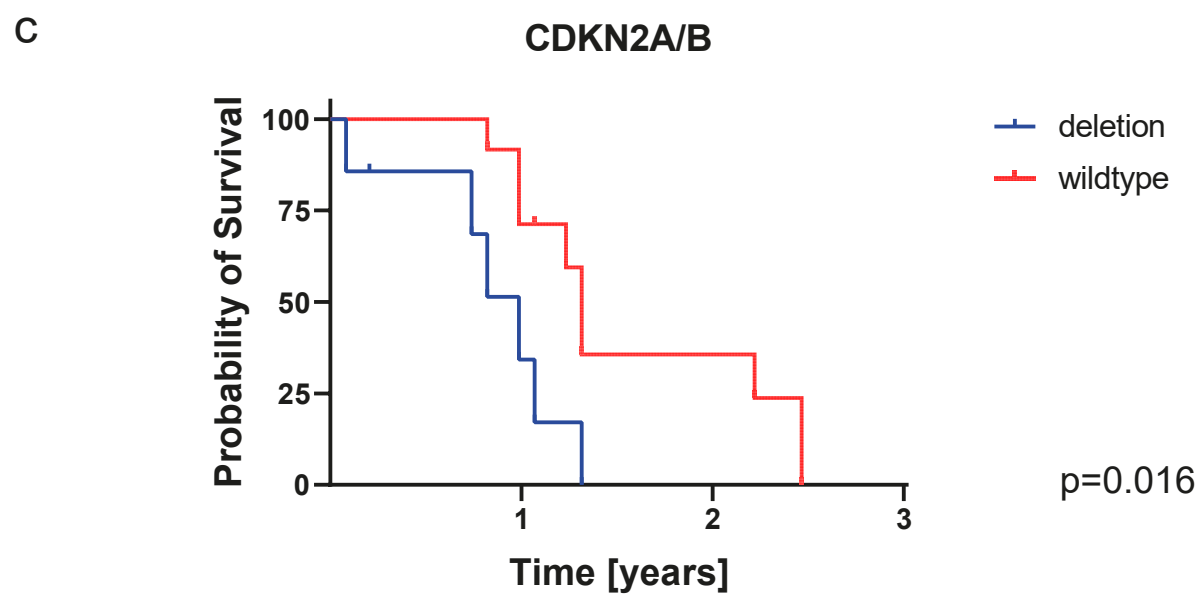
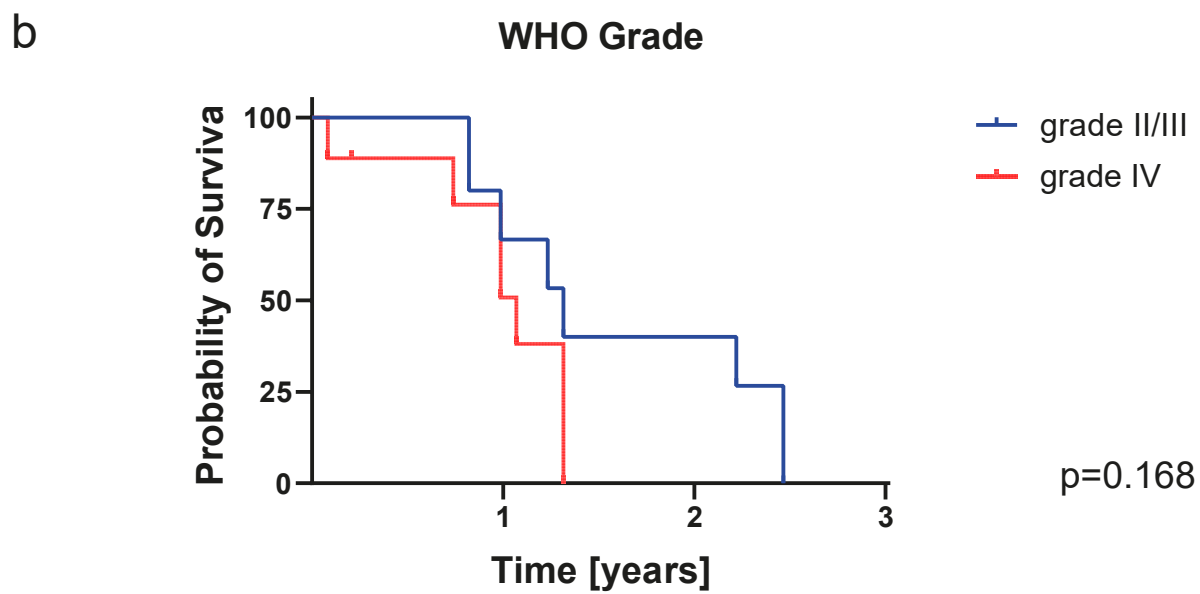
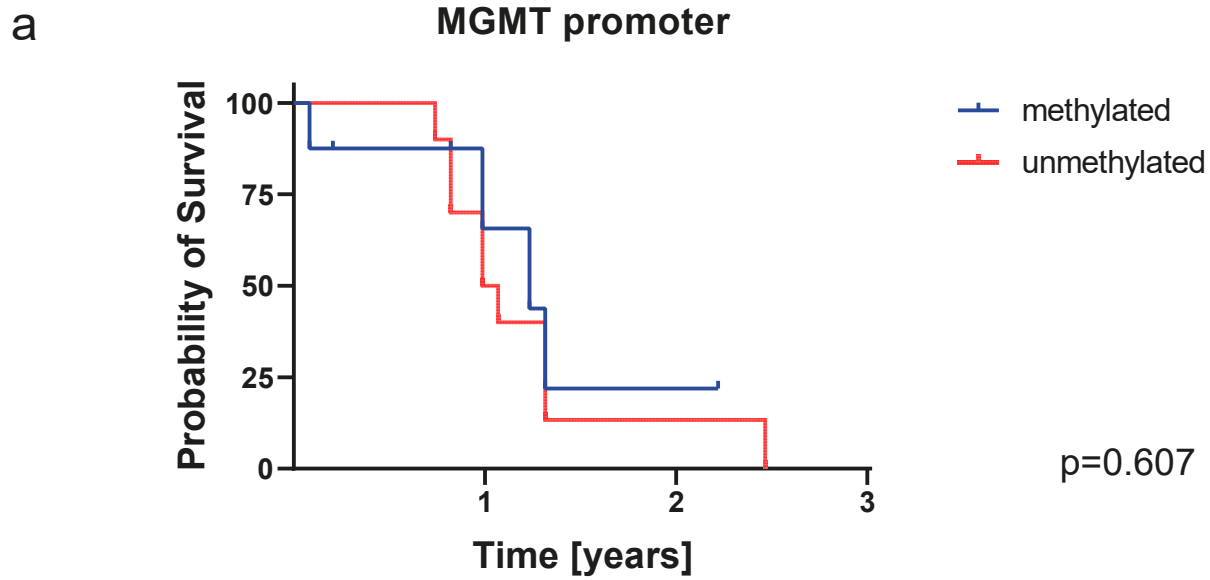
Mean methylation values among IDH-mutant gliomas. Beta-values vary from 0 (unmethylated) to 1 (fully methylated).



No. at risk

| | | | | | | | | |
|---|----|---|---|---|---|---|---|---|
| — | 19 | 3 | 0 | 0 | 0 | 0 | 0 | 0 |
| — | 24 | 8 | 3 | 3 | 3 | 1 | 1 | 1 |

IDH-wt glioma data was taken from
Guerrini-Rousseau et al., 2019 (PMID:32642664)



Supplementary Table 1: Treatment of PMMRDIA

| No | Mut. | Syndrome | P/R | OS | PFS | Death | MGMT | Loc. | Diagn. | Grade | Treatment |
|----|------|----------|-----|----|-----|-------|---------|------|--------|-------|---------------------------------|
| 1 | MSH6 | cMMRD | P | 10 | 10 | no | meth | H | EPN | III | |
| 2 | MLH1 | | P | 27 | 3 | yes | not det | H | astro | III | IR + TMZ (Stupp) |
| 3 | MSH6 | cMMRD | P | 1 | 1 | yes | meth | H | GBM | IV | |
| 4 | MSH6 | | P | 15 | 10 | yes | meth | H | oligo | III | IR + TMZ (Stupp), CCNU, Avastin |
| 5 | MSH2 | | U | | | yes | unmeth | H | PNET | IV | |
| 6 | MSH6 | Lynch | P | 12 | 8 | yes | unmeth | H | GBM | IV | IR + TMZ (Stupp), TTF, IR |
| 7 | MSH6 | cMMRD | P | 12 | 11 | yes | meth | H | GBM | IV | IR, Nivolumab |
| 8 | MSH6 | cMMRD | P | | | | unmeth | H | GBM | IV | |
| 9 | MSH6 | | U | | | | meth | | HGG | | |
| 10 | | | P | 16 | 10 | yes | unmeth | | GBM | IV | only resection |
| 11 | | cMMRD | P | 16 | 10 | yes | meth | H | GBM | IV | IR, TMZ |
| 12 | MSH6 | | U | | | | unmeth | | HGG | | |
| 13 | MSH6 | cMMRD | P | 13 | 13 | no | unmeth | H | GBM | IV | Nivolumab |
| 14 | MSH2 | Lynch | P | 12 | 6 | yes | unmeth | H | astro | II | IR, TMZ, Pembrolizumab |
| 15 | MSH2 | Lynch | P | 6 | | no | unmeth | H | GBM | IV | IR, TMZ, CCNU |
| 16 | MSH6 | | P | 16 | 11 | yes | unmeth | H | astro | III | IR, TMZ |
| 17 | | | R | 27 | 27 | no | meth | H | GBM | IV | IR + TMZ (Stupp), CCNU |
| 18 | | | P | 30 | 10 | yes | unmeth | H | astro | III | IR + TMZ (Stupp), CCNU, Avastin |
| 19 | MSH6 | cMMRD | P | 10 | | yes | unmeth | H | astro | III | |
| 20 | MLH1 | Lynch | P | 10 | | no | meth | H | astro | III | |
| 21 | MSH6 | cMMRD | P | 13 | | yes | unmeth | PF | GBM | IV | |
| 22 | MSH6 | Lynch | P | 2 | 2 | no | meth | H | GBM | IV | IR, CCNU |
| 23 | | | P | 10 | | yes | unmeth | H | astro | III | IR, TMZ |
| 24 | | | P | | | yes | unmeth | H | GBM | IV | |
| 25 | | | P | 9 | 9 | yes | unmeth | | GBM | IV | TMZ |
| 26 | | | U | | | | unmeth | | | | |
| 27 | | | U | | | | meth | | | | |
| 28 | | | U | | | | unmeth | | | | |
| 29 | | | U | | | | meth | | | | |
| 30 | | | U | | | | meth | | | | |
| 31 | | | U | | | | unmeth | | | | |
| 32 | | | U | | | | unmeth | | | | |