**Table 1**

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Reference | Phase | Pt # | MedianAge, (range) | CR/PR | RR | TMZ(mg/m2) | Irinotecan(mg/m2)xdxw | Otheragents |
| Wagner (PBC,2007)[1] | R | 16 | 18 (7-33) | 1/3 | 25% | 100 x 5 | IV 10-20 x 5 x 2 | None |
| Casey (PBC 2009)[2]  | R | 19 | 19.5 (2-40) | 5/7 | 63% | 100 x 5 | IV 20 x 5 x 2 | None |
| Hernandez-Marques (An Ped 2013)[3] | R | 8 | 13 (6-18) | 0/3 | 37% | 80-100 x 5 | IV 10-20 x 5 x 2 | None |
| Raciborska (PBC 2013) [4] | R | 22 | 14.3 | 5/7 | 54% | 125 x 5 | IV 50 x 5 | VCR |
| McNall-Knapp (PBC 2010)[5] | I | 1 | NA | 0/1 | 100% | 100 x 5 | IV 20 x 5 x 2 | VCR |
| Wagner (PBC 2010)[6] | I | 5 | (<21) | 1/1 | 40% | 100-150 x 5 | PO 35-90 x 5 | VCR |
| Wagner (PBC 2013)[7] | I | 2 | 20,22 | 1/1 | 100% | 150 x 5 | PO 90 x 5 | VCR, BEV |
| Bagatell (PBC 2014)[8] | I | 7 | (<21) | 0/1 | 14% | 100-150 x 5 | PO 50-90 x 5 | TMS |
| Kurucu (Ped Hem Onc 2015)[9] | R | 20 | 14 (1-18) | \*see text | 55% | 100 x 5 | IV 20 x 5 x 2 | None |
| Anderson (Exp Opin 2008)[10] | R | 25 | 15 | 7/9 | 64% | 100 x 5 | IV 10 x 5 x 2 | None |
| Palmerini (Acta Onc 2018)[11] | R | 51 | 21 (3-65) | 5/12 | 34% | 100 x 5 | IV 40 x 5 | None |
| Salah (Clin Transl Oncol 2021)[12] | R | 45\* | 20 (5-45) | 1/11 | 27% | 100 x 5 | IV 40 x 5 in 21 ptIV 50 x 5 in 24 ptIV 20 x 5 x 2 in 6 pts | None |
| McCabe (Proceedings ASCO)[13] | II | 88 | 19 (4-49) | 4/14 | 20% | 100 x 5 | IV 50 x 5 | None |

R: retrospective; I: Phase I; II: Phase 2; IV: intravenous; PO: oral; TMZ: temozolomide; VCR: vincristine; BEV: bevacizumab; TMS: temsirolimus; Uk: unknown; NA: not applicable; PBC: Pediatric Blood and Cancer; An Ped: Annals of Pediatrics; Ped Hem Onc: Pediatric Hematology and Oncology; Exp Opin: Expert Opinion Investigational Drugs; Acta Onc: Acta Oncologica; Clin Trans Oncol: Clinical and Translational Oncology;\*see text

**Total experience 312 patients, 111 Objective responses, Objective response rate: 36%**

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11. Palmerini, E., et al., *Irinotecan and temozolomide in recurrent Ewing sarcoma: an analysis in 51 adult and pediatric patients.* Acta Oncol, 2018. **57**(7): p. 958-964.

12. Salah, S., et al., *Irinotecan and temozolomide chemotherapy in paediatric and adult populations with relapsed Ewing Sarcoma.* Clin Transl Oncol, 2021. **23**(4): p. 757-763.

13. McCabe, M.G., et al., *Results of the second interim assessment of rEECur, an international randomized controlled trial of chemotherapy for the treatment of recurrent and primary refractory Ewing sarcoma (RR-ES).* Journal of Clinical Oncology, 2020. **38**(15\_suppl): p. 11502-11502.