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A multimodal strategy based on surgery, radiotherapy, ICE regimen and high dose chemotherapy in atypical teratoid/rhabdoid tumours: a single institution experience.

[Fidani P](#), [De Ioris MA](#), [Serra A](#), [De Sio L](#), [Ilari I](#), [Cozza R](#), [Boldrini R](#), [Milano GM](#), [Garrè ML](#), [Donfrancesco A](#).

Division of Pediatric Oncology Unit, Ospedale Pediatrico Bambino Gesù, Piazza S. Onofrio 4, Rome, 00100, Italy. fidani@opbg.net

PURPOSE: Atypical Teratoid/Rhabdoid Tumour is a rare and aggressive childhood tumour. The outcome of a series treated with the same multimodal strategy was reported. **PATIENTS:** The patients were treated with surgery, 2 courses of ifosfamide/carboplatin/etoposide(ICE), 2 courses of cyclophosphamide/etoposide/carboplatino/thiotepa (CECAT) or 2 other ICE courses, high dose chemotherapy (HDC) and radiotherapy. **RESULTS:** Eight patients underwent primary surgery achieving a complete removal in 3. Progressive disease (PD) occurred in 2/8 patients during ICE courses and in 3/4 during CECAT courses. After 4 courses 5 patients presented a PD. HDC was performed in 3 patients followed by local radiotherapy. The Kaplan Meier OS and EFS probability at 5 years are, respectively, 50% (CI 11-80%) and 33% (CI 6-66%). **CONCLUSION:** A strategy based on surgery, including a second surgical look, and on radiotherapy appears the best option. ICE regimen and HDC correlate with good prognosis in some patients but this approach needs further evaluation.

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