

RESEARCH ARTICLE



Outcome and prognostic factors in high-risk childhood adrenocortical carcinomas: A report from the European Cooperative Study Group on Pediatric Rare Tumors (EXPeRT)

Giovanni Cecchetto¹ | Alba Ganarin¹ | Ewa Bien² | Peter Vorwerk³ |
 Gianni Bisogno⁴ | Jan Godzinski⁵ | Patrizia Dall'Igna¹ | Yves Reguerre⁶ |
 Dominik Schneider⁷ | Laurence Brugières⁸ | Pierre Leblond⁹ | Andrea Ferrari¹⁰ |
 Ines Brecht¹¹ | Angela De Paoli¹² | Daniel Orbach¹³

Pediatr Blood Cancer 2017; 64: e26368

- 82 patients <18 years (study period 2000-2013) from databases of Germany, France, Poland and Italy
- 62 localized disease, 20 with metastases
- Whole serie: 3-year PFS 39%, OS 55%
- Localized diseases: 3-year PFS 51%, OS 73%
- Metastatic diseases: 1-year PFS 6%, OS 29%

PFS and OS were influenced by distant metastases, tumor volume, lymph node involvement, age, presence of two or more risk factors, vascular involvement (OS) and incomplete surgery (OS).

At multivariable analysis, the main factors at PFS were volume > 200 cm³ (HR 2.6) and distant metastases (HR 8.26). OS was significantly influenced by the presence of metastases (P < 0.0001)

Conclusions:

Distant metastases and large tumor volume were the main unfavorable prognostic factors. Presence of two or more factors related to ACC was associated with an aggressive behavior of disease.

