

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 Cecchetto<sup>1</sup> | Alba Ganarin<sup>1</sup> | Ewa Bien<sup>2</sup> | Peter Vorwerk<sup>3</sup> |  
 Gianni Bisogno<sup>4</sup> | Jan Godzinski<sup>5</sup> | Patrizia Dall'Igna<sup>1</sup> | Yves Reguerre<sup>6</sup> |  
 Dominik Schneider<sup>7</sup> | Laurence Brugières<sup>8</sup> | Pierre Leblond<sup>9</sup> | Andrea Ferrari<sup>10</sup> |  
 Ines Brecht<sup>11</sup> | Angela De Paoli<sup>12</sup> | Daniel Orbach<sup>13</sup>

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- 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 cm<sup>3</sup> (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.

