

## MANAGEMENT OF PATIENTS WITH ADRENOCORTICAL CARCINOMA

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The prognosis of patients with adrenocortical carcinoma (ACC) is still poor. Although most diagnostic and therapeutic strategies are not evaluated according to criteria of evidence-based medicine, most experts usually agree in general on most “standard” diagnostic work-up and treatment options. However, even these recommendations of expert groups are often not yet translated into clinical practice. The main obstacle for optimal practice is the rarity of the disease with an incidence of only 1-2/million/year. Furthermore, ACC represents a heterogeneous group of neoplasia which makes it difficult to predict outcome and response to treatment. In general, patients with ACC should be considered for surgical resection which is feasible in all patients with stage I-II and most patients with stage III disease. In the majority of cases, adjuvant treatment with mitotane is considered necessary and in some cases, radiotherapy can be helpful to decrease the rate of local recurrence. In those patients who are diagnosed at a time point when metastatic spread has already taken place systemic treatment options include chemotherapy in combination with mitotane. Currently, drug regimens including etoposide, doxorubicin and cisplatin versus streptozotocin have been tested in the first randomized trial on ACC patients. Due to improved international networking ([www.ensat.org](http://www.ensat.org)), further clinical trials have become reality including testing of an IGF1 receptor antagonist in stage IV ACC patients and evaluation of adjuvant mitotane in low risk patients after complete resection of ACC.