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Valassi, E.[a b] , Castinetti, F.[c] , Ferriere, A.[d] , Tsagarakis, S.[e] , Feelders, R.A.[f] , Netea-Maier, R.T.[g] , Droste, M.[h] , Strasburger, C.J.[i] , Maiter, D.[j] , Kastelan, D.[k] , Chanson, P.[l] , Webb, S.M.[a m] , Demtröder, F.[n] , Pirags, V.[o] , Chabre, O.[p] , Franz, H.[q] , Santos, A.[a m] , Reincke, M.[r]

Corticotroph tumor progression after bilateral adrenalectomy: data from ERCUSYN
(2022) *Endocrine-Related Cancer*, 29 (12), pp. 681-691.

DOI: 10.1530/ERC-22-0074

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Abstract
Corticotroph tumor progression after bilateral adrenalectomy/Nelson's syndrome (CTP-BADX/NS) is a severe complication of bilateral adrenalectomy (BADX). The aim of our study was to investigate the prevalence, presentation and outcome of CTP-BADX/NS in patients with Cushing's disease (CD) included in the European Registry on Cushing's Syndrome (ERCUSYN). We examined data on 1045 CD patients and identified 85 (8%) who underwent BADX. Of these, 73 (86%) had follow-up data available. The median duration of follow-up since BADX to the last visit/death was 7 years (IQR 2–9 years). Thirty-three patients (45%) experienced CTP-BADX/NS after 3 years (1.5–6) since BADX. Cumulative progression-free survival was 73% at 3 years, 66% at 5 years and 46% at 10 years. CTP-BADX/NS patients more frequently had a visible tumor at diagnosis of CD than patients without CTP-BADX/NS ($P < 0.05$). Twenty-seven CTP-BADX/NS patients underwent surgery, 48% radiotherapy and 27% received medical therapy. The median time since diagnosis of CTP-BADX/NS to the last follow-up visit was 2 years (IQR, 1–5). Control of tumor progression was not achieved in 16 of 33 (48%) patients, of whom 8 (50%) died after a mean of 4 years. Maximum adenoma size at diagnosis of CD was associated with further tumor growth in CTP-BADX/NS despite treatment ($P = 0.033$). Diagnosis of CTP-BADX/NS, older age, greater UFC levels at diagnosis of CD and initial treatment predicted mortality. In conclusion, CTP-BADX/NS was reported in 45% of the ERCUSYN patients who underwent BADX, and control of tumor growth was reached in half of them. Future studies are needed to establish effective strategies for prevention and treatment. © 2022 Society for Endocrinology Published by Bioscientifica Ltd.

Author Keywords
bilateral adrenalectomy; corticotroph adenoma; Cushing's disease; Nelson's syndrome

Index Keywords
bevacizumab, cabergoline, corticotropin, pasireotide, temozolomide, cytidine triphosphate; abducens nerve paralysis, ACTH secreting cell, adult, adverse outcome, Article, bilateral adrenalectomy, cancer control, cause of death, clinical feature, cohort analysis, controlled study, cranial nerve paralysis, Cushing disease, diabetes insipidus, early intervention, female, follow up, growth hormone deficiency, hormone determination, human, hydrocortisone blood level, hyperpigmentation, hypogonadism, hypophysis adenoma, hypopituitarism, hypothyroidism, macroadenoma, major clinical study, male, microadenoma, mortality, Nelson syndrome, ophthalmoplegia, opticus nerve palsy, outcome assessment, patient monitoring, pharmaceutical care, postoperative complication, prevalence, progression free survival, radiation dose fractionation, radiosurgery, risk factor, tumor growth, tumor volume, adrenalectomy, adverse event, Cushing disease, Nelson syndrome; Adrenalectomy, Corticotrophs, Cytidine Triphosphate, Humans, Nelson Syndrome, Pituitary ACTH Hypersecretion

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Publisher: BioScientifica Ltd.

ISSN: 13510088
CODEN: ERCAE
PubMed ID: 36197784
Language of Original Document: English
Abbreviated Source Title: Endocr.-Relat. Cancer
2-s2.0-85141888729
Document Type: Article
Publication Stage: Final

Source: Scopus

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