the corticotropin test. Seventy-three (41%, CI95%: 34-48%) patients had an insufficient response to the corticotropin test. Neither the risk of adrenal insufficiency, unstimulated nor stimulated P-cortisol levels were directly associated with any of the GR SNPs. However, for both insensitive SNPs 9β and ER23/23EK the effect of current prednisolone dose on stimulated P-cortisol was smaller (higher dose did not suppress the cortisol level as much) in carriers vs. non-carriers (p=0.035 and p=0.0075). The same sensitivity-associated tendency was seen for the N363S, but not the *Bcl*1 SNP. The *Bcl*1 SNP occurred more frequently in our cohort compared with control groups (63% vs. 40%, p<0.0001). The same trend was seen for the other sensitive but less frequent SNP N363S. The 9β SNP also occurred more frequently in our cohort (18% vs. 13%, p=0.029), but depending on regional sub cohorts in one control group.
CONCLUSION: The GR SNPs did not directly associate to the risk of adrenal insufficiency, unstimulated nor stimulated cortisol levels, respectively. However, the effect of prednisolone dose on stimulated cortisol depended on the GR SNPs: Cortisol was less suppressed with higher current prednisolone dose in patients carrying the insensitive SNPs. The substantially higher frequency of the *Bcl*1 SNP is remarkable even with modest n=239. It questions whether there is an association between carrying the sensitive GR SNPs and inability to taper GC treatment ending up in this cohort of long-term treated patients.

Pediatric Endocrinology

PEDIATRIC OBESITY, THYROID, AND CANCER

Risk of Long-Term Endocrine Sequelae in Survivors of Progressing Childhood Optic Pathway Glioma Treated by Upfront Chemotherapy: Preliminary Analyses of 102 Subjects from the French Multicentric BB-SFOP Registry

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For the brain tumor committee of SFCE (Societe´ Française des Cancers de l'Enfant). Objective: Therapeutic approach favors chemotherapy as the first-line-treatment in progressing OPG. There are few data on long term endocrine outcomes of aggressive OPG treated by upfront chemotherapy. Our main objective was to describe the long-term endocrine sequelae in these patients and to identify potential early predictors of the endocrine involvement. Subjects and methods: Children diagnosed with OPG at an age younger than 16 years from the French multicentric BBSFOP registry were included. They were treated with upfront chemotherapy according to the BB-SFOP protocol in France between June 1990 and December 2004, and subsequent treatment (second-line chemotherapy, surgery, radiotherapy) was used depending on tumor progression. They underwent a late evaluation with clinical and biological assessment between January 2011 and March 2016. Results: One hundred and two patients were included in our study. The mean age at tumor diagnosis was 3.3±0.3 years. The mean time of follow-up was 13.9±3.7 years. A history of precocious puberty was present in 36% of the subjects. At least one endocrine deficiency was present in 93% of the subjects (GHD 74%, TSH deficiency 57%, ACTH deficiency 36%, hypogonadotropism 33%, gonadic deficiency 30%, diabetes insipidus 15%; inappropriate AVP secretion 7%). 37% of males and 39% of females were overweight or obese. Mean adult height, reached in 51 subjects, was -1.2±1.3 SDS in males, and -0.7±1.4 SDS in females. Chemotherapy only was protective from pituitary deficiencies (odds ratio 0.19 to 0.37, $P < 0.05$). NF1 was protective from TSH and ACTH deficiencies (odds ratio 0.25 to 0.35, $P < 0.05$). Tumor volume on diagnostic MRI was not predictive of pituitary deficiencies. Gonadic deficiency was significantly more frequent in males than females (46,5% vs 12.2%, $P < 0.05$), and associated with chemotherapy only (OR 3.2, $P < 0.05$) and NF1 (OR 4.8, $P < 0.05$). Overweight/Obesity was associated with ACTH deficiency (OR 5, $P < 0.05$).Conclusion: Obesity and late endocrine dysfunction were frequent in subjects treated by upfront chemotherapy for aggressive OPG during childhood. However, chemotherapy only, when possible, was protective from pituitary involvement.

Neuroendocrinology and Pituitary

NEUROENDOCRINOLOGY AND PITUITARY

Systematic Screening Reveals Large Number of Undiagnosed and Untreated Cardiovascular Risk Factors in Adults with Prader-Willi Syndrome

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Introduction: Prader-Willi syndrome (PWS) is a complex hypothalamic disorder, combining hypotonia, intellectual disability (ID), pituitary hormone deficiencies and hyperphagia. In PWS, up to 3% of patients die every year. In half of the patients, the cause of death is obesity related and / or of cardiovascular (CV) origin.
Obesity is caused by hyperphagia combined with a low energy expenditure. Untreated hormone deficiencies like hypogonadism and hypothyroidism can cause low muscle mass and low basal rest metabolism (BRM) leading to this low energy expenditure. Patients with PWS should exercise one hour daily to compensate for their low BRM. However, hormone deficiencies usually cause fatigue, leading to exercise intolerance. Musculoskeletal and / or behavioral problems can also cause reduced physical activity. The subsequent sedentary lifestyle can induce CV risk factors like hypertension, hypercholesterolemia and diabetes mellitus (DM).
Another risk factor often present in PWS is sleep apnea, which can be central (CSA), obstructive (OSA) or both. Both CSA and OSA can lead to pulmonary hypertension and a further increase in obesity.
The above mentioned health problems often remain unnoticed and untreated, which is partly due to the behavioral phenotype of PWS (patients seldomly report pain and hardly ever complain about physical problems). However, if left untreated, these risk factors can cause CV complications leading to hospital admission or even death. To reveal yet