The aim of this study was to evaluate clinical and molecular aspects of a Brazilian cohort of pediatric patients with PPGLs. Out of 262 patients with PPGLs, 26 (9 %) were diagnosed before 19 yrs of age (16 males and 10 females), with a median age of 14.5 yrs (range, 4 to 18). Genetic investigation was performed in 19 patients: 14 by automated Sanger sequencing (*VHL, SDHB, SDHD* and *RET* genes) and 5 by a custom next-generation sequencing (NGS) panel including all genes previously associated with germline mutations in PPGLs. Median tumor size was 5.5 cm (1.7 to 16). Pheochromocytomas (PHEOs), paragangliomas (PGLs) or both were diagnosed in 46%, 31% and 23% of the patients, respectively. Bilateral PHEOs were diagnosed in 61% of the cases, most of them asynchronous (75%). Genetic diagnosis was confirmed in 14 out of 19 (74%) patients and all variants were found in heterozygous state: 8 *VHL* missense mutations from 6 kindreds (p.R167W in 2 kindreds, p.R167Q in one and p.G114S in 3); 3 *SDHB* mutations (p.C98Y, c.201-2A>G and p.L180L); 2 *SDHD* mutations (p.Y144_H145del and p.Q121*); and one *RET* mutation (p.C634R). All 8 VHL patients had bilateral PHEOs and 3 of them had also abdominal PGLs. All patients with *SDHB* mutations had abdominal PGLs. Two patients with *SDHD* mutation had head and neck paraganglioma (one of them had unilateral PHEO). Genetic investigation by NGS Panel was negative in all 5 cases: 2 malignant PPGLs (one PHEO and one PGL) and 3 PHEOs. Four out of 26 (15%) pediatric PPGLs were malignant: 2 with *SDHB* mutation and 2 with negative screening (one PHEO and one PGL). In conclusion, the majority of pediatric PPGLs (74%) were hereditary and almost exclusively caused by mutations in hypoxic genes. *VHL* (PHEOs) and *SDHB* (only PGLs) were the most frequent affected genes in this cohort of pediatric PPGLs.
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Reproductive Endocrinology

MALE REPRODUCTIVE HEALTH - FROM HORMONES TO GAMETES

Adoption of an Age Adjusted Testosterone Reference Range Reduces Referrals to Endocrine Clinic and New Prescriptions of Testosterone

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Testosterone levels decline with age. However, until recently well defined harmonised age and/or obesity (BMI <30kg/m^2) adjusted reference ranges did not exist.[1] There is also a lack of international consensus on whether an age adjusted reference range (RR) should be used to define the syndrome of hypogonadism in men. Our local referral guideline suggests referral to endocrinology is appropriate if morning testosterone is <9.4nmol/L similar to the Endocrine Society Clinical Practice Guideline.[2] In mid 2018 our laboratory adopted the published *all men* age adjusted RR[1]. We sought to; i) investigate clinic referrals before and after adoption of the *all men* age adjusted RR and, ii) to model the impact on referrals and prescription of testosterone replacement therapy (TRT) had we adopted either the lower limit of either *all men* or *non-obese* age adjusted RR as our referral criteria. Despite similar numbers of testosterone levels being measured in the laboratory, referrals to endocrine clinic for investigation of male hypogonadism fell by 52% (n=101 vs 48) in the one year following the introduction of the new age adjusted RR, with a corresponding reduction in prescriptions for testosterone. Mean testosterone concentration (6.7±2.5 vs 6.4±3.9nmol/L [mean±SD], NS), and age (51±13.9 vs 50±17.9 years, NS) of individuals referred were similar before and after the change of RR. Of the 101 patients referred for investigation of hypogonadism prior to the new RR mean testosterone concentrations were 8.5±4.5, 7.3±4.1, 6.8±3.6, 6.7±2.1 & 6.6±1.6nmol/L, with 39, 71, 39, 40 & 17% of the 87 patients seen in clinic being prescribed TRT in age groups 19-39 (n=28), 40-49 (n=7), 50-59 (n=33), 60-69 (n=20) &70-79 (n=6) respectively, excluding those with a history of anabolic steroid use or Klinefelter's syndrome. Mean BMI was 30.9±4.4kg/m^2, which was similar between age groups. Had the lower limit of normal of the *all men* testosterone RR been employed as our referral criteria in the preceding year, 23.8% (24/101) of referrals would not have met referral criteria, and 26.2% (n=11/42) of those receiving a prescription would potentially not have received a trial of TRT. In contrast, had the *non-obese* age adjusted RR had been adopted for all men 13.9% (14/101) of referrals would not have met referral criteria and, of those prescribed testosterone, 2.4% (n= 1/42) would not have received a trial of TRT. In conclusion, adoption of the *all men* age adjusted RR for testosterone has been associated with a significant fall in referrals for investigation of male hypogonadism. However, modelling of historical clinic data would suggest that some non-obese individuals miss out on a therapeutic trial of TRT, especially if the *all men*, rather than *non-obese*, age adjusted RR is adopted.

Reference: (1) Travison et al, J Clin Endocrinol Metab, 2017,102(4):1161-1173, (2) Bhasin S et al,. J Clin Endocrinol Metab. March 2018;103(5):1715-1744.

Neuroendocrinology and Pituitary

PITUITARY TUMORS II

Long-Acting SSA Treatment Patterns in Sweden From 2005 to 2017: A Nationwide Study

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Background

Acromegaly is a complex disease, primarily treated with pituitary surgery or long-acting somatostatin analogues (LA-SSA). Few studies have examined real-world use of LA-SSA. This analysis evaluated LA-SSA treatment patterns in Sweden for patients with acromegaly.

Methods