

with essential hypertension (EH) diagnosed by West China Hospital from 2009 to 2018 were retrospectively analyzed. The diagnostic performance of different screening methods was compared by plotting receiver operating characteristic curves (ROC). **Results:** The proportion of low potassium in PA group was higher than that in the EH group (86.20% vs. 36.50% $P < 0.001$). The area under the ROC curve (AUC) of upright ARR was greater than that of upright AA2R, upright PRA, upright PAC, supine ARR, and lowest blood potassium ($P < 0.05$); The AUC of logistic regression model (nomogram) [which consists of the upright PAC, the upright PRA, and the lowest blood potassium] was greater than that of upright ARR (96.3% vs. 94.6%, $P < 0.05$). **Conclusion:** The upright ARR is the best single screening indicator; AA2R (radioimmunoassay) is not recommended for PA screening. Ultimately, the logistic regression model (nomogram) is superior to the upright ARR.

Pediatric Endocrinology

PEDIATRIC SEXUAL DIFFERENTIATION, PUBERTY, AND BONE BIOLOGY

Understanding and Communication Around DSD According to the Mothers and Patients' Perspectives

LIA MESQUITA LOUSADA QUINTAO, MD¹,
SORAHIA DOMENICE, PhD², Elaine Maria Frade Costa,
MD, PHD³, Tania A. Bacheega, MD, PHD⁴,
Eveline Gadelha Pereira Fontenele, PhD⁵,
Debora Cabral Coutinho, MD⁶, Rafael Loch Batista, MD PhD⁷,
Maria Tereza Martins Ferrari, MD¹,
Berenice Bilharinho Mendonca, MD⁸.

¹Hospital das Clinicas - FMUSP, Sao Paulo SP, Brazil, ²Disciplina de Endocrinologia e Metabologia, Laboratorio de Hormônios e Genética Molecular/LIM42, Hospit, So Paulo, Brazil, ³University of Sao Paulo, Sao Paulo, Brazil, ⁴So Paulo University, Sao Paulo, Brazil, ⁵Universidade Federal do Ceara, Fortaleza, Brazil, ⁶HC-FMUSP, Sao Paulo, Brazil, ⁷Hospital Das Clinicas, Sao Paulo SP, Brazil, ⁸Univ Sao Paulo Fac Med, Sao Paulo, Brazil.

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Understanding and Communication around Differences of Sex Development (DSD) according to the mothers and patients' perspectives

Communication around DSD is complex. It involves diagnosis and treatment aspects and is influenced by the psychological status of the individuals and the cultural context. An adequate understanding by patients and relatives is essential for approach of DSD. Objective: To evaluate the DSD care setting in three Brazilian tertiary centers in order to identify the barriers to an adequate understanding and an optimal communication. Methods: A guide with 69 questions, assessing the level of knowledge, the main doubts and difficulties around DSD was developed and guided individual interviews with 100 mothers and 53 adult patients with DSD. The main doubts were clarified and a self-assessment was requested to them before and after the interview on a scale from 0 to 10. Results: Mothers and patients mean age were, respectively, 35.2 and 36.5 years. Both of them had a satisfactory educational level. Although 48% ($p < 0.01$) of mothers and 68% ($p < 0.02$) of patients were satisfied/very satisfied about their knowledge related to the DSD, 78% and 58%, respectively, of them still had doubts. The doubts

were related to diagnosis, karyotype, medications, appearance of genitalia, surgery, sexual activity, fertility, genetic counseling, consequences of the condition and treatment on general health and condition influence on the child's behavior and personality. The unsatisfied mothers cited as barriers to an optimal understanding the complexity of the conditions, the difficult terms and the psychological stress at diagnosis. Patients also cited as barriers the absence of dialogue about the condition at home and some of them chose not to know. About 55% of mothers and 62% of patients didn't even know the name of the condition; but positively 88% of them knew the necessary treatment. Regarding communication, 68% of mothers and 89% of patients didn't feel comfortable talking to people about the DSD condition and around 68% of them underwent negative comments. Although 73% of patients would prefer to be first informed about their condition at childhood, 29% of mothers think that childhood is the best age for it. Among mothers and patients, the most and least appropriate term to name the DSD condition is, respectively, genital malformation and disease ($p < 0.01$). Both of them have the stigma as the main concern. Conclusion: Even in a tertiary center with a multidisciplinary team, the mothers and patient's knowledge about DSD conditions is scarce. The proper choice of the term to refer to DSD conditions should consider the families and patients perspectives. Communication about DSD is prejudiced by the lack of knowledge and the stigma suffered by these patients and families. Thus, due to complexity of this topic, continued educational action must be instituted as a strategy to modify this scenario.

Neuroendocrinology and Pituitary

ADVANCES IN NEUROENDOCRINOLOGY

Deficient Fear Extinction in PRKARIA-Defective Mice

MARGARET KEIL, PhD¹, Enrica Paradiso, PhD¹, Rita S. Keil, BS², Maddalena Ugolini, MS¹, Evan Harris, BS¹, T. John Wu, PhD², Constantine A. Stratakis, MD, D(med)Sci, PhD(hc)¹.

¹NIH/NICHD, BETHESDA, MD, USA, ²USUHS, BETHESDA, MD, USA.

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Background: The role of the cAMP/PKA signaling in molecular pathways involved in fear memory is well established: PKA is required for fear memory formation and is a constraint for fear extinction. Previously we reported that a *Prkar1a* heterozygote (HZ) mouse that was developed in our lab to investigate Carney complex (CNC), the disease caused by *PRKARIA* mutations, showed brain region-specific increased PKA activity that was associated with anxiety-like behavioral phenotype and threat bias (Keil, 2010, 2013). We hypothesized that *Prkar1a*^{+/-} (HZ) mice would have deficits in fear extinction behavior. Brain derived neurotrophic factor (BDNF) has a critical role in formation of fear memory and its transcription is regulated by PKA/CREB. A mouse model with down regulation of PKA provides an opportunity for the first time to investigate the effect of altered PKA signaling on fear conditioning and extinction.

Method: Fear conditioning, fear extinction learning, and fear extinction recall were tested in adult male HZ and

wild-type (WT) mice as follows: fear conditioning training followed 24hr later by extinction training (new context), then 24hr later by extinction recall training. Percentage of time freezing was used to assess conditioned fear response. We measured BDNF gene expression in brain regions after completion of extinction recall training.

Results: As expected, fear conditioning (learning) behavior was similar in HZ and WT mice. However, HZ mice showed a significant deficit in the early phase of fear extinction learning compared to WT. There was no difference in extinction recall between genotypes. Alterations in BDNF gene expression in the prefrontal cortex and amygdala was associated with deficit in fear extinction.

Conclusion: Mice with a downregulation of *Prkar1a* gene demonstrate intact fear conditioning but impaired fear extinction learning, consistent with prior studies that report that PKA inhibition is necessary to facilitate extinction learning. *Prkar1a*^{+/-} mice provide a valuable model to investigate impaired fear extinction to identify mechanisms for therapeutic targets for anxiety and trauma-related disorders.

Adrenal

ADRENAL CASE REPORTS II

Adrenal Cortical Carcinoma in a Male Patient Who Presented with Classic Signs and Symptoms of Cushing's Syndrome

Jeffrey Magnaye Humarang, Doctor of Medicine.

MAKATI MEDICAL CENTER, Manila, Philippines.

SUN-156

Adrenal Cortical Carcinoma in a Male Patient who Presented with Classic Signs and Symptoms of Cushing's Syndrome

Background: Adrenal Cortical Cancer (ACC) is a rare malignancy with an annual incidence of 1-2 per million population. ACC is generally considered a highly malignant tumor and account for 0.05-2% of all cancer. Clinical case: A 32-year old male, newly diagnosed diabetic and hypertensive for 1 month, with a BMI of 37.91 kg/m², presented with intermittent, vague, non-radiating flank pain. Within a month, patient started to develop weight gain associated with Cushingoid fascies, buffalo hump, elevated blood sugars, hypertension, and eventually developed purple striae in the abdomen associated with easy bruisability. Ultrasound was done which showed bilateral nephrolithiasis. Further work-up with CT-stonogram was done which showed bilateral nephrolithiasis and a 55x37x60mm heterogeneous mass above the left kidney in the area of the left adrenal gland. Further imaging with whole abdominal CT scan was done which revealed stable size of the mass in the region of the left adrenal gland with heterogeneous attenuation and enhancement. Initial hormonal work-ups done showed abnormal 1mg dexamethasone suppression test (874mmol/L N: 172-497mmol/L), but with normal aldosterone (73.22, N: <90pg/ml), renin (8.60pg/mL, N: 5-40pg/mL) and 24-hour urine metanephrines (31 mcg/24 hours N: 115-695mcg/24 hours). Serum K and blood glucose was managed accordingly and patient was cleared to undergo surgery. Patient underwent left unilateral laparoscopic adrenalectomy

which revealed a 7cm soft, friable left adrenal mass with minimal hematoma on the antero-inferior portion of the adrenal gland, with active bleeding. Histopathology of the left adrenal mass revealed high-grade adrenal neoplasm, consistent with adrenocortical carcinoma. Postoperatively, patient did not have complications of hypo nor hyperglycemia, hypotension and serum potassium was stable. Hydrocortisone was given postoperatively in tapering doses. Repeat cortisol were done 24 and 48 hours post op which showed normal results (4.02ug/dL and 5.37 ug/dL respectively, N: 5-25 ug/dL). Patient was referred to Oncology service for Chemotherapy and was eventually discharged stable with home medications for his Diabetes mellitus. On follow-up, there was noted improvement on the signs and symptoms of Cushing's syndrome, with no hypokalemia and better blood glucose control. Conclusion: Majority of adrenal mass are benign but a high index of suspicion for malignancy should always be part of the workup. The acuteness of symptoms and size of mass are important determinants of malignancy. Prior to adrenalectomy, perioperative endocrine therapy is a must. Adrenocortical carcinoma should be managed by a multidisciplinary specialist team including an endocrinologist, oncologist, surgeon, radiologist, and a histopathologist.

Neuroendocrinology and Pituitary

NEUROENDOCRINOLOGY AND PITUITARY

A Pilot Study: Comparing Prolactin Measurements Between Two Different Immunoassays

Walter K. Woznick, MD¹, Almira Yang, DO², Ian Gunsolus, PhD¹, Jenna Lynne Sarvaideo, DO³.

¹Medical College of Wisconsin, Germantown, WI, USA, ²Medical College of Wisconsin, Brookfield, WI, USA, ³Medical College of Wisconsin, Whitefish Bay, WI, USA.

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Abstract: Hyperprolactinemia from a prolactin-secreting pituitary tumor is the most common endocrine disorder of the hypothalamic-pituitary axis. As suggested in the 2011 Endocrine Society Guidelines on Diagnosis and Treatment of Hyperprolactinemia, macroprolactin level should be assessed in patients with asymptomatic hyperprolactinemia. However, as discussed in prior studies comparing the performance of common prolactin immunoassays in a reference population of both males and females with and without known hyperprolactinemia or macroprolactinemia, there has been poor harmonization between assays and variable reactivity towards macroprolactin, resulting in significantly different normal ranges for total and monomeric prolactin between manufacturers. The goal of our analysis is to assess the concordance of the Roche and Siemens prolactin immunoassays using cases in which prolactin and macroprolactin testing was ordered on clinical indication. We hope to educate clinicians regarding potential variability between assays that may not be fully accounted for by using established, assay-specific reference ranges. We reviewed patients 18 years and older from any gender who underwent evaluation of prolactin levels as clinically indicated and had elevated serum prolactin on a Roche assay with a subsequent normal prolactin on a Siemens assay. Seven