Pediatric Adrenocortical Tumors (PACT)
A single tertiary center experience: Clinical, Biological and Pathological Characteristics Analysis

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Background
PACT are primary tumors of the adrenal cortex, with heterogeneous presentation and varied behaviors in terms of progression. Generally diagnosed by clinical manifestations linked to hormone excess. They are more frequent in females with maximal incidence between 1-4 years of age. Complete tumor resection is the only intervention that provides the best chance for cure. Efficacy of adjuvant treatment in advanced disease (stages III and IV) is undetermined.

Aim
To evaluate the demographic, clinical, biochemical, and pathologic characteristics in a cohort of PACT in a single tertiary institution of Argentina.

Subjects and Methods
Retrospective review of 28 medical records of PACT (chronological age (CA) <18 years (y)) treated between 1987-2017. Data analyzed were: Demographical, clinical (Height-SDS, BMI-SDS, and bone age (BA), biochemical (serum DHEAS levels), and histological features (Wieneke index1). Staging (ST) according to COG system2, and therapeutic interventions (surgery and adjuvant chemotherapy, Achemo).

Results
Demographical distribution of patients

Clinical complaint at diagnosis
- Hypertensive Encephalopathy: 7% (n=2)
- Abdominal mass/pain: 56% (n=10)
- Virilization: 57% (n=16)
- Hormonal Overproduction: 45% (n=13)

Mean duration of symptoms was 10.9 months.

Biochemical and Pathological features according to COG staging

Distribution by age and sex
- CA, years (median 2.8y)
  - < 5: 21 (75%)
  - 5 - 12: 4 (14.3%)
  - 13 - 18: 3 (10.7%)
- Sex
  - Female: 20 (71.4%)
  - Male: 8 (28.5%)

Antropometric features according hormonal predominance

Clinical Features
- Virilization: 16 (57.1%)
- Cushing’s Syndrome: 10 (35.7%)
- Mixed: 2 (7.1%)

Antropometric features:
- Height-SDS
- BMI-SDS
- BA/CA

Overall (OS) and Disease Free Survival (DFS) according to COG staging
- ST I
- ST II
- ST III
- ST IV

Overall and Disease Free Survival according to AChemo

Conclusions
We reported the experience in our cohort of 28 PACT seen in a single center of Argentina over 30 years. Height-SDS and BMI-SDS mirror ACT hormonal secretion. Very high serum DHEAS levels might be use as a biological marker of tumor stage. Less advanced disease were associated with best patient outcomes. Long term follow-up is needed to draw valid conclusions of using AChemo.

References: