165 TESTICULAR TUMORS WITH PEUTZ-JEGHERS SYNDROME (PJS). Darrell M. Wilson, William C. Pitts, Raymond L. Hintz & Ron G. Rosenfeld. Stanford University, Departments of Pediatrics & Pathology, Stanford, CA. USA.

Females with PJS (mucocutaneous pigmentation and intestinal polyposis) have an increased risk of ovarian tumors, with 5% developing a pathologically distinct neoplasm described as a sex cord tumor with annular tubules. We evaluated a 6 yr old male with PJS for bilateral gynecomastia. Testes both measured 3 ml and were normal to palpation. Although two initial estradiol levels were less than 10 pg/ml, a subsequent value was moderately elevated at 24 pg/ml. T4, FSH, LH, T, prolactin and DHAS were all within the normal range for his age. Bone age was slightly advanced at 8.2 yr. Abdominal and pelvic ultrasound, including both adrenals, was normal. Ultrasonography of the testes, however, demonstrated multiple echogenic areas in both testes. Open biopsy revealed multiple minute foci of large-cell, calcifying Sertoli cell tumors. Since the lesions did not appear to be malignant by histology, and were bilateral, the testes were not removed. Subsequently, there has been no progression of the gynecomastia, and serum estradiol has remained <20 pg/ml. Although similar tumors have been reported in two other prepubertal males with PJS, this case is unique in that the tumors were 1) bilateral and multifocal, 2) nonpalpable, and 3) detected by ultrasound. Taken together, these reports strongly suggest that males, as well as females, with PJS are at risk for gonadal tumors and, as such, all patients with PJS should be closely followed for the potential development of gonadal tumors.

166 THE ASSESSMENT OF DIMINISHED TESTICULAR FUNCTION IN PUBERTAL AGED BOYS: IS HCG TESTING NECESSARY? Howard E. Kulin and Steven J. Santner. The M.S. Hershey Medical Center of The PA State University, Depts. of Pediatrics and Medicine, Hershey, PA 17033 U.S.A.

The evaluation of anorchia or diminished testicular reserve usually involves hCG testing. We questioned whether such a study is useful in the presence of the pubertal gonadotropin rise since endogenously elevated LH (and FSH) may provide an adequate stimulation test. Consequently, we serially measured urinary gonadotropin and serum testosterone (T) in 9 anorchic boys, ages 10-17, and 8 individuals, ages 9-18, with diminished testicular function. Nine of these patients also received hCG stimulation tests. Twenty normal boys, ages 10-17, and 34 adult men furnished urine samples for comparative purposes. By age 12 the anorchic boys excreted adult amounts of LH (>500 mIU/hr) in the presence of low serum T (<33 ng/dl); hCG stimulation tests in 4 of these individuals revealed no increments in T. The LH increments in patients with diminished gonadal reserve were similar to those found in the anorchic boys; T values, however, ranged from 90-640 ng/dl with post-hCG increments of 107-1200 ng/dl. An early FSH rise served to separate patients with decreased testicular function from normals since FSH attained adult levels (>200 mIU/hr) by age 11 and castrate levels (>1700 mIU/hr) by age 13. In conclusion, urinary gonadotropin and serum T measurements can be used to define the gonadal status of boys by age 12; hCG testing is not required in the early pubertal period to distinguish anorchia from diminished testicular reserve.

167 ZINC STATUS IN PATIENTS WITH KALLMANN SYNDROME AND ITS RESPONSE TO TESTOSTERONE TREATMENT. Mariano Castro-Magaña, Moris Angulo, Shang Yao Chen, Platon Jack Collipp, Ajanta Derenoncourt and Boris Espinoza. Nassau County Med Ctr, SUNY, Stony Brook Health Sci Ctr, Dept of Ped., E. Meadow, NY 11554.

The observation that oral testosterone (T) administration raises hair zinc (Zn) levels in children with constitutional growth delay has led us to study the hair, serum, and urine Zn concentrations before and after the intramuscular administration of T enanthate (300 mg/month) in 7 male children (15-18y) with Kallmann Syndrome (KS) (Table).

	Before T ($\bar{x}$ ± SD)	After T ($\bar{x}$ ± SD)	P Value
Hair Zn(ug/g)	119.9 ± 15.4	183.3 ± 11.6	<0.001
Urine Zn(ug/g Creat)	1000.6 ± 355.4	423.3 ± 152.3	<0.001
Serum Zn(ug/dl)	94.9 ± 12.9	113.8 ± 10.1	<0.02

We have shown that growth hormone (GH) administration to hypopituitary children resulted in significant reduction in urinary Zn excretion and increase in hair Zn levels. Now we have found that male adolescents with KS are also Zn deficient and that T administration resulted in a remarkable reduction in the urinary excretion of Zn and significant increase in their hair and serum Zn levels. These observations support the concept that anabolic hormones like GH and T play an important role in Zn metabolism.

168 FEMALE PSEUDOHERMAPHRODITISM CAUSED BY THE TRANSPLACENTAL TRANSFER OF TESTOSTERONE OF OVARIAN ORIGIN. Val Abbassi and David M. Gibbons, Department of Pediatrics, Georgetown University School of Medicine, Washington, D.C.

Fetal masculinization of maternal ovarian origin is usually caused by androgen secreting tumors. The case of a severely masculinized female infant described here is believed to have been caused by transplacental transfer of maternal testosterone probably secreted by polycystic ovaries. Ovulation was induced by Clomid. Hirsutism and cystic acne developed during pregnancy. Baby was delivered at term by C-section. Polycystic ovaries were observed at operation. Other than ambiguous external genitalia the infant was healthy. Scrotum was small but well formed. Phallus measured 20 mm with a small opening at the base. No gonads were palpable. Genitogram showed a urethral tract with no evidence of a vagina. Karyotype showed a normal (46 XX) chromosomes. Cord testosterone was 1232 ng/dl. Cord and postnatal serum 17α-OH progesterone and urinary 17 KS, 17 KGS were all normal. Maternal testosterone at 9 days post-partum was 468 ng/dl and declined to 82 ng/dl at 18 days. Free testosterone remained elevated. 2000 HCG x 3 days did not increase testosterone. Prednisone failed to modify total and free testosterone. However, cyclic estrogen and progesterone lowered total and free testosterone to 10 ng/dl and 1.6 pg/ml respectively. At laparotomy the infant had normal ovaries, fallopian tubes and uterus, but no vaginal structure could be identified.

The extreme masculinization in this infant is probably caused by transplacental transfer of testosterone produced by polycystic ovaries under intense HCG stimulation.

169 J.M.Cuatrecasas-Membrado, L.Audi-Parera, A.Zuasnabar, M.Catalá, A.Puig, J.M.Bosch-Banyeras (Intr.by E.Vicens) Department of Pediatrics, Hospital General Granollers Hormonal Laboratory, Hospital Badalona (Catalonia) SPAIN.

A CASE OF MALE PSEUDOHERMAPHRODITISM WITH XYY KARYOTYPE

The association between a female phenotype and XYY Karyotype is a very rare ocurrence. We report the third case of male pseudohermaphroditism with female genitalia, morphologically normal testes and XYY Karyotype described so far to our knowledge.

A 3 year old girl was admitted to our department for bilateral inguinal hernia repair. Physical examination was unremarcable. Two testis were identified with a seminiferous tubules diameter of 78.0 ±12.4um (M ±SD) witch corresponded to 7-8y. controls, absence of spermatogonia and interstitium showing some hyperplastic Leydig cell nests. No uterus or anexa could be seen and the vagina was 4 cm long, ending into a blind pouch. Peripheral lymphocytes and skin fibroblasts showed a 47,XYY Karyotype. Two stimulation tests with hCG (1500 UI/48h.x7) were carried out measuring testosterone (T), dihydrotestosterone (DHT), androstendione, 17 OHprogestrone, progesterone and dehydroepiandrosterone plasma levels. The plasma T response to hCG was sligthly below the normal range (231 ng/100ml, controls 624±249). Baseline and hCG stimulated T precursors were appropriate for age and the T/DHT ratio under hCG was normal. LH and FSH reponses to LHRH stimulation were increased. DHT receptors in genital skin fibroblasts (GSF) presented normal binding capacity (20.2 fmol/mg prot., controls=26.5±8.3) and normal affinity. 5-alpha-reductase activity in GSF was very low (1.6 pmol / mg prot/h versus 177.6±76.5 in controls).

The complete lack of virilization in this patient suggests an androgen insentivity with normal DHT receptor concentration in GSF and secondary 5-alpha-reductase deficiency associated to a 47,XYY Karyotype.

170 MENSTRUAL DYSFUNCTION IN ADOLESCENTS WITH INCREASED INTRACRANIAL PRESSURE. Geoffrey P. Redmond, Gita Gidwani, Janet Bay, Joseph Hahn, David Rothner, Depts of Endocrinology, Gynecology, Neurological Surgery, Neurology and Pediatrics, Cleveland Clinic Foundation, Cleveland, Ohio USA

The menstrual cycle is controlled by the central nervous system and alterations in its function may be a reflection of neurological disease. Although longstanding increased intracranial pressure (ICP) may produce subtle but important changes such as learning disabilities, there may be few clues to identify increased ICP as the cause. We have recently seen two patients with longstanding learning disabilities in whom amenorrhea provided the impetus for CT scanning. One patient presented at 20 years of age and had an initial prolactin of 42. She had a 3rd ventricle cyst causing obstructive hydrocephalus. The second patient presented at 17 years of age with amenorrhea and severe headaches. Prolactin was 29.5 ng/ml. Hydrocephalus due to a 4th ventricle ependymoma was found on CT. Spontaneous menses occurred in the second patient after placement of a shunt. Her prolactin level gradually fell to normal but menses resumed before the prolactin fall. The first patient initially required medroxyprogesterone acetate postoperatively but resumed spontaneous menses when she was placed on spironolactone for her hirsuitism. CONCLUSIONS: 1) Amenorrhea may be an important clue to the presence of increased intracranial pressure and intracranial neoplasia, 2) Hypothalamic compression rather than hyperprolactinemia appears to be the mechanism of amenorrhea in our cases.