family renamed the child "Brenda." Unaware of her history, Brenda struggled with significant gender identity, psychological, and behavioral issues throughout her childhood and adolescence. When made aware of this history, she transitioned to male gender and assumed the name "David." After years of psychological distress, David Reimer committed suicide in 2004. Despite the myriad lessons gleaned from this tragic story, medical and surgical management of children with atypical genitalia still remains often misguided, as providers continue to assume paternalistic roles in determining sex assignment and surgical interventions. A fifteen year old XY male with Robinow Syndrome presented for evaluation of hypogonadism and urinary incontinence. At birth, the patient was discovered to have a micropenis and perineal hypospadias and was diagnosed with hypogonadotropic hypogonadism. At the recommendation of the medical team, the infant underwent bilateral orchiectomy at eight months of age followed by urethroplasty and vaginoplasty at six years of age. The child was then given a female sex assignment. At twelve years of age, the child felt discordant from the sex of rearing and wished to be identified as male—his natal, genetic sex. He transitioned to male gender and began testosterone injections. He had history of recurrent UTIs and severe incontinence requiring diaper use. He strongly desired neophallus and urethral reconstruction for improved quality of life. The patient endorsed prior depression and desires to self-harm. He had significant concerns regarding his gender presentation and transition. He shared his difficulties in continuing in the same school system with peers who knew him as a female prior to transition and was concerned about peers knowing his medical history. In the years since the famous David Reimer case, the medical system has made tremendous strides in recognizing the need for patient autonomy and shared decision-making in patients with Differences of Sex Development and genital atypia. However, the paternalistic history of this field continues to leave its indelible mark more than 20 years since David Reimer's case made headlines, as physicians continue to recommend definitive sex assignments and surgical interventions. As with the David Reimer case, the bodily integrity of this XY infant was altered in a permanent fashion with inadequate education of his family and little to no credence given to the autonomy of the child himself. We, as physicians, cannot continue to paternalistically apply John Money's concept of gender neutrality and rigidly mandate sex assignments and early surgical interventions.

Pediatric Endocrinology

PEDIATRIC ENDOCRINOLOGY CASE REPORT

Pediatric Giant Prolactinoma Presenting With Acute Obstructive Hydrocephalus and Intracranial Hypertension

Grace Hendrix, MD[1], Robert Benjamin, MD[2], Nancie J. MacIver, MD,PhD[2], Daniel P. Barboriak, MD[2], Pinar Gumus Balikcioglu, MD[3].

[1]Duke University Hospital Endocrine Fellowship Program, Durham, NC, USA, [2]Duke University Medical Center, Durham, NC, USA, [3]Duke University Medical Center, DURHAM, NC, USA.

Background: Pediatric prolactinomas (PP) are rare but represent 50% of all pediatric pituitary adenomas. Girls are affected more frequently than boys, although PP tend to be larger and more aggressive (earlier age, larger mass, and higher prolactin levels) in boys. Thus, microadenomas (tumors < 10 mm in diameter) are typical in females and macroadenomas (10–40 mm in diameter) are typical in males. Giant prolactinomas (> 40 mm in maximum diameter), an unusual subset of macroprolactinomas, are also commonly found in boys. In a large case series, the largest tumor volume reported was 93.5 cm^3. Here we report a giant prolactinoma in a female requiring V/P shunt for decompression. **Clinical Case:** A 16-year old female presented with 2 weeks of intractable headache, nausea and vomiting, vision impairment, and changes in balance described as running into stationary household objects. Historical review revealed primary amenorrhea and short stature. On initial exam, the patient had a right eye afferent pupillary defect, concern for loss of color vision, and bilateral optic nerve edema with blurred disc margins. Brain MRI showed a large lobulated mass centered in the suprasellar cistern, measuring approximately 6.4 x 5.8 x 5.7 cm with a tumor volume of 105 cm^3. There was extension superiorly, anteriorly, and laterally, with homogeneously enhancing and cystic components, and mass effect resulting in obstructive hydrocephalus. Differential diagnoses included craniopharyngioma, germinoma, and adenoma. Initial tests demonstrated prolactin of >2,000 ng/mL, with diluted result of 17,811.16 ng/mL. Morning fasting labs confirmed multiple anterior pituitary hormone deficiencies including central hypothyroidism, ACTH deficiency, GH deficiency, and hypogonadotropic hypogonadism. The patient was started on hydrocortisone and levothyroxine. Due to obstructive hydrocephalus and vision impairment, she underwent VP shunt placement for decompression. She was started on cabergoline for medical treatment of the tumor and did not require surgical resection. Repeat prolactin measurements have shown striking improvement (to 2,350 ng/ml, 824 ng/ml, and 152 ng/ml at 1 week, 1-month, and 2-month-follow-up, respectively) with central vision improved in both eyes, papilledema resolved, and resolution of headaches. **Conclusion:** Giant prolactinomas presenting with hydrocephalus and intracranial hypertension are very rare in pediatrics, especially in girls, and can vary greatly in mass characteristics and resulting hormone deficiencies. Our patient is unique with her large tumor volume and the extent of pituitary hormone deficiencies. Prolactin levels should be measured with all sellar masses, as this may prevent unnecessary invasive intervention and possibly provide prompt response to medical management.

Pediatric Endocrinology

PEDIATRIC ENDOCRINOLOGY CASE REPORT

Pituitary Macroprolactinoma Apoplexy in a Prepubertal Girl

Sabliny Carreiro Ribeiro, MD[1], Angélica Cristina Dall agnese, MD[1], Laís Marques Mota, Md[2], Cesar Geremia, MD[1], Marina Bressiani, MD[1], Michele Teixeira Hertz, MD[1], Lucas Bandeira Marchesan, MD[2], Márcia Khaled Coutinho, PhD[1].

[1]Hospital Criança Conceição, Porto Alegre, Brazil, [2]Hospital Nossa Senhora da Conceição, Porto Alegre, Brazil.