Fall, 2011 Volume 1, Issue 1





Greetings from the Children's Hospital at Downstate Medical Center and Long Island College Hospital:

I hope that you enjoy the first edition of our new quarterly bulletin for Pediatricians, along with the late summer weather. As many of you already know, after more than an eleven year hiatus I have returned to Downstate. While I have always remained an active Pediatric Gastroenterologist, I look forward to spending even more time devoted to patient care. However, I also hope to serve as your personal connection to the medical school and our two hospital campuses, Downstate Medical Center and Long Island College hospital. Our chair, Dr Stanley Fisher and the entire faculty are committed to providing the best possible health outcomes for the children in the borough and this region. This can only be attained by creating long standing partnerships with the primary care providers throughout the area. We pledge to be available and accountable to you and your practice.

As the demands on Pediatricians get more difficult, we will be exploring novel ways in which the Children's Hospital can help you provide care and keep up with issues that matter to your practice. Let me know what topics you would like to hear about in our weekly grand rounds and if you would prefer evening programs. Maybe you have encountered an unusual case and would like to know what tests should be ordered even before you refer to a sub-specialist. Perhaps you are wondering what you should be doing to help your overweight children? Or else you may just feel that your children with sickle cell disease or asthma could just be doing better. If you would like to email me specific questions, I can forward your inquiries to the appropriate individual. If a visit to your office is more convenient we could arrange for that. Lastly, if an evening or early morning meeting with a number of your peers would work best we can help organize and coordinate that.

In future editions, we will have other members of the department discuss what they can offer to you, your staff, and your patients. Again, we are open to finding out what you need so let us know. In this edition, we will include a challenging case from the Children's Hospital with some commentary from one of our sub-specialists. Lastly, we will include a listing of important future events at the medical school. Have a great fall season, Simon.

Em Da

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Autism and Autistic Spectrum Disorder (ASD) affects many families in Brooklyn, New York. Please join us Sunday, September 25, 2011 as we build an integrated, unified network of programs, services and supports for young people with ASD.

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Every edition, we will choose a challenging patient case to include in the newsletter: Rather than describing a condition that is extremely rare, we will seek out cases that could be a part of your busy pediatric patient practice. Each will highlight an unusual or unanticipated nuance of the condition. We will also have an expert from our faculty provide you with an insightful discussion. Please let me know what you think and feel free to email me potential cases for future editions.

A lean, muscular, athletic 13yr old (122lb, 5'5") female was seen for worsening acanthosis nigricans (AN). There were prominent raised hyperpigmented patches in both axille. There was only one velvety patch on her left posterior neck with areas of minimal hyperpigmentation and minimal hirsutism. With the epidemic of obesity upon us, all pediatricians have become very familiar with AN and the risk of diabetes and the metabolic syndrome, however, what should the pediatrician consider in the nonobese adolescent? The discussion of this case will be by

Dr. Elka Dickman-Jacobson MD.



Polycystic Ovary Syndrome by Dr. Elka Dickman-Jacobson, MD

Acanthosis nigricans (AN) is a physical finding that is often associated with insulin resistance. Although Pediatricians typically think of AN has being associated with type II diabetes. There are other entities that can present with the typical discoloration, velvety skin most prominent in the posterior neck and axillae. One of the more commonly encountered conditions is Polycystic ovary syndrome (PCOS), including "lean PCOS" as the girl in the vignette. PCOS accounts for the vast majority of anovulatory symptoms and hyperandrogenism in adolescent girls.

PCOS is not a disease, but rather a syndrome arising from multiple etiologies with variable clinical presentations. Predominant characteristics include: menstrual irregularity (oligo-or amenorrhea, or irregular bleeding), cutaneous hyperandrogenism (hirsutism, severe acne, male pattern alopecia), polycystic ovaries on ultrasound, obesity and insulin resistance. Obesity contributes to the insulin resistance of PCOS, however insulin resistance is disproportionately elevated compared to the degree of adiposity. Furthermore, 40-50% of girls with PCOS are lean.

PCOS patients have exaggerated ovarian androgen production. This may be related to insulin resistance, hypothalamic-pituitary-ovarian dysregulation and adipose tissue dysfunction. Dysregulation of local follicle regulatory systems by androgens and other factors impedes normal follicular growth, resulting in follicular arrest at the 4 to 8 mm diameter size, and normal ovulation does not occur

PCOS is clinically heterogeneous and diffcult to diagnose. Girls with PCOS may present with any one of the following-hirsutism, menstrual irregularities, obesity, acanthosis nigricans, premature pubarche, and/or precocious puberty. AN may be the presenting complaint of patients with PCOS. In addition to those with obvious signs of insulin resistance and obesity, other high risk groups include patients with diabetes mellitus (type 1, type 2, or gestational), a history of premature adrenarche, having a first-degree relatives with PCOS, or those using anti-seizure medication (valproic acid appears to be the major culprit).

An array of endocrine studies should be conducted on girls with biochemically confirmed hyperandrogenism in order to exclude other diagnoses prior to initiating therapy, regardless of whether the ultrasound is normal or consistent with PCOS. This list includes: serum prolactin, insulin-like growth factor-I (IGF-I), thyroid function, 17-hydroxyprogesterone, cortisol levels, and bhCG in amenorrheic patients. In patients with PCOS, these studies will be normal. Any abnormal result suggests another cause of hyperandrogenism, and must be further evaluated.

The treatment of PCOS should be tailored to the prevailing signs and symptoms. The combination oral contraceptive pill (OCP), which contains estrogen and progestin, has traditionally been the first-line treatment for patients with PCOS and menstrual irregularity, especially in patients with cutaneous signs of androgen excess. The treatment of obesity improves ovulation, acanthosis nigricans, androgen excess, and cardiovascular risk in patients with PCOS. Diet and exercise are the first-line treatment for obese adolescents with PCOS. Metformin has been shown to attenuate both insulin resistance and androgen excess and has an essential role in the management of metabolic and reproductive abnormalities in PCOS. Thiazolidinediones and sulfonlyureas have serious safety issues and are therefore not recommended for PCOS. Masking the presence of excess hair and physical hair removal are cornerstones in treating hirsutism; however, costs are not covered by third-party payers.

While PCOS was once considered an aesthetic and/or fertility disorder, it is now recognized to confer substantial morbidity, particularly in the spectrum of glucose regulatory abnormalities and Type 2 Diabetes Mellitus. Other long term associations include cardiovascular disease, obstetrical complications such as spontaneous abortions, and mood disorders. The combination of the high prevelance and serious prognosis of PCOS should lower our threshold for evaluating for this syndrome, even in cases with a paucity of symptoms. Prompt recognition and treatment of PCOS can have a significant impact on prognosis including increasing fertility, improving psychological well being, and prolonging life.

Two telephone number for referrals to Pediatric Sub-specialists of the Children's Hospital at Downstate: (Downstate Medical Center & LICH)

(718)270-4714 (Downstate) *(718)780-1025 (L1CH)

Adolescent Medicine

Amy Suss, MD Rhonda Cambridge, MD Kim Dumont-Forrester MD (LICH)*

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Haesoon, Lee, MD Madu Rao, MD Stephen Wadowski, MD

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Urology

Mark Horowitz, MD

Sleep Disorders Center

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- 4. AmeriChoice
- 5. AmeriGroup
- Atlantis Health Plan 6.
- 7. CarePlus
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- Community Care Partners-Center Care
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- 20. Galaxy Health Network
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- 38. NY Medicare CMS 855R
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Sleep Medicine Specialists for Adults and Children

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The **Children's Digestive Health Center** offers state-of-the-art facilities for: Diagnostic and therapeutic upper GI, endoscopy and colonoscopy, Esophageal and anorectal manometry, Esophageal pH monitoring, Gastrostomy tube placement, Hydrogen breath testing, Liver biopsy and Video capsule endoscopy.

Appointments may be scheduled at locations in central Brooklyn, Brooklyn Heights, Staten Island and Manhattan, close to public transportation with off-street parking at all sites. Our offices are open Mon-Fri, 8:30 AM -5:00 PM, with most appointments scheduled within 1 week (immediate appointments are always available for urgent problems). For questions or to arrange a visit, please call one of our offices:

Brooklyn & Staten Island: (718) 270-8884 (general questions)

Brooklyn & Staten Island: 718) 270-4714 (appointments)

> Manhattan: (212) 844-8300 (appointments)

Pediatric
Gastroenterologist
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The Children's Digestive Health
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