Disorders of Sexual Differentiation (DSD)
A Consensus Statement

A committee appointed by Arne Ljungqvist, MD, Chair of the International Olympic Committee Medical Commission has focused on what action current science would recommend if an athlete were diagnosed with a disorder of sexual differentiation (DSD). Their concern was with the elite athlete and whether some disorders might provide a woman with an unfair advantage in competition. The definition of "unfair" remains to be determined. While gender tests for female athletes are no longer a requirement for participation in women's events, testing for performance enhancing drugs and methods remains part of the effort to promote fair play in Olympic competition. Unfortunately, the biochemical effect of some medical conditions may result in testosterone levels well beyond the normal range for women. If so, these women run the risk of being unfairly identified as potential users of a performance enhancing drug. Early identification of these conditions would benefit the athletes by allowing corrective action of the medical problem at a younger age, sparing them the trauma of being investigated for illegal use of drugs and being denied participation in Olympic events.

WomenSport International in cooperation with the American College of Sports Medicine recently convened a Consensus Conference to discuss this issue (June 2010, Baltimore, USA). The results of our deliberations are outlined below.

Definition of a female:

An individual identified as female at birth and who has been raised and lives as a female.

Individuals with a Y chromosome, and individuals with no Y chromosome but with a condition that provides an endogenous hormonal performance advantage, may still be considered female but will be subject to rules, regulations and testing regarding competition.

An individual identified as male at birth who subsequently has undergone surgery for gender reassignment is identified as a transgender woman. Recommendations regarding competition as a transgender female are already in place within the IOC for these women.

Consensus Points:

1. All women have the right to participate in sport

2. Every woman should be treated with dignity and concern for her physical and mental health.

3 Organizations doing any testing of athletes must have a strict confidentiality protocol in place, one that has been approved by their sports governing body and must be followed explicitly.

4. Educational information regarding DSD should be distributed to Sports Medicine Associations, International and National Sports Governing bodies, High School Athletic Associations, Team Physician courses, etc.

5. An educational program directed at the public, media, coaches, athletes and sports administration regarding DSD should be developed. Information on prevalence and potential health implications should be included.

6. A valid algorithm for the detection and diagnosis of DSD in female athletes needs to be developed and validated for eventual implementation in sport practice. Published procedures for diagnosis should include evidence of false positive rates for diagnostic tests to avoid the devastating psychological consequences of misdiagnosis.

7. A standardized Pre-Participation Health Examination (PPHE) should be implemented by sport governing bodies for all participating athletes. The PPHE should facilitate the early detection of numerous serious and treatable medical conditions.

8. All sports medicine physicians should be educated about and aware of DSD in sport to ensure the identification of the condition during the PPHE.

9. Psychological counselling should be made available for any athlete identified with a DSD.

10. Collaboration with the IOC working group on DSD should be encouraged by having a qualified representative or representatives of WSI available to attend their future meetings.

The Pre-participation Health Exam (PPHE)

The Pre-participation Health Exam (PPHE) is a tool to be encouraged for all sport participants to screen for potential health concerns in athletes involved in sport. The focus of the PPHE should be on the maintenance of athlete health and be carried out in a sensitive, respectful manner. It may identify athletes with DSD. Athletes identified with potential concerns regarding sexual development on screening with the PPHE should be referred for further investigation and treatment if necessary to an appropriately qualified specialist.

Athletes with a DSD should only be identified by medical personnel as part of the PPHE. They should not be subjected to testing for DSD on the basis of challenges from competitors, coaches or anyone else solely on the basis of "masculine" appearance, outstanding performance, etc.

Centers of excellence to assist athletes with DSD should be made available for diagnostic and treatment purposes. These centers should address the physical and psychological implications of DSD. These centers should be accessible geographically and financially.

Given the wide variability and heterogeneity of people with DSD, eligibility for competition should be determined on a case by case basis. Well developed policies and procedures with strict confidentiality will ensure a humane and fair process for those identified athletes. Shifting the focus on DSD to one of health through education will serve to protect the athlete from the psychological trauma of inappropriate media exposure and public scrutiny.

Competition Standards:

An individual without a Y chromosome who falls within standardized testing levels may compete in a women's division. An individual born and raised as a female, regardless of karyotype, who has undergone treatment so as to abrogate any advantage of endogenous hormonal performance enhancement, may compete in a women's division.

Disorders of Sexual Development (DSD)

Appendix 1

Androgen Excess Conditions

Definition: Any condition in which a woman has a level of androgens above the normal range for females. These conditions are estimated at 8% prevalence in the US and most likely have a similar prevalence in other countries.

ENDOGENOUS SOURCES: Androgens that are made within the body.

Polycystic Ovary Syndrome (PCOS): PCOS is the most frequently encountered condition of androgen excess with the ovaries producing higher than normal amounts of estrogen, progesterone, and testosterone. Usually the levels are not excessively high, and would not normally test out of range by drug testing agencies. PCOS occurs in approximately 5-10% of women, and is associated with acne, hirsutism, and infertility (also coronary heart disease, diabetes, and metabolic syndrome). Fifty percent of women with PCOS are overweight. This condition does not often lead to ambiguous or virilized genitalia. It is not classified as a DSD, but is the most common "mild" androgenic disorder.

Congenital Adrenal Hyperplasia (CAH): CAH is a genetic condition that results in abnormal steroidogenesis. Different types of CAH include excesses/deficits of mineralocorticoids or excesses of androgens. Excessive androgens can cause virilization of the female.

Androgen Insensitivity Syndrome (AIS): AIS is a genetic condition in a XY individual in which receptors do not recognize male hormones, or androgens. AIS can be complete or incomplete, thus presenting with varying degrees of undervirilization.

Tumors: Tumors of the ovary (rare), and adrenal gland (rare) produce excess testosterone. These tumors have up to 30% malignant potential, or chance of becoming cancerous. Examples of an ovarian tumor: Sertoli-Leydig/ Leydig, Lipoid, Granulosa-theca cell, teratoma, gonadoblastoma, and Hilus cell tumor. Examples of adrenal gland tumors: adenoma, carcinoma.

Pregnancy: In pregnancy, testosterone increases to term. Virilization of the mother or a female infant can occur if an exaggerated response to increased human chorionic gonadotropin (HCG) occurs in the ovarian stroma (luteoma). It has been postulated that the physiologic changes that occur naturally in pregnancy such as an increase in Hb/Hct, VO2 max, plasma volume etc, could also enhance performance in certain sports.

Differential Diagnosis: Other increased androgen conditions of androgen excess include Cushing's syndrome, hypothyroidism, HAIR-AN syndrome (hyper androgen- insulin resistance- acanthosis nigricans), hyperprolactinemia, Idiopathic hyperandrogenism, and PCOS. Increased precursor androgen can be seen in liver disease and in stress (by increased adrenal androstenedione).

Testosterone- a steroid hormone produced by testes, ovaries and the adrenal gland. It has anabolic activity, meaning it increases protein synthesis within cells (anabolism), especially in muscles. It also has a virilizing, or androgenic effect. Most circulating testosterone is bound to SHBG, sex hormone binding globulin, Androgenicity is dependent upon the unbound fraction, which enters the cell and activates the receptors. .

EXOGENOUS SOURCES:

Medications: Elevated androgens can also be from exogenous sources:

Examples:

Oral Contraceptives - certain birth control pills have androgenic-acting hormones present

Danocrine (danazol)- used for treatment of endometriosis

Testosterone and related substances- can be prescribed for decreased libido

Testosterone in petrolatum- used topically to treat a vulvar condition called lichen sclerosis

Anabolic androgenic steroids (AAS)- these drugs simulate testosterone and dihydrotestosterone, and are used to stimulate bone growth and to increase appetite. They can treat chronic wasting conditions, such as cancer and AIDS. Examples are oral methyltestosterone and injectable dianabol.

Normal ranges- women

Total testosterone: 6-86 ng/dl, suspicious for tumor= >200ng/dl
Free (Unbound) testosterone: 0.7-3.6 pg/ml
Dehydroepiandrosterone sulfate (DHEAS): 35-40 ug/dl
Testosterone/epitestosterone (T:E) ratio: < 4
Androstenedione: 0.7-3.1 ng/dl
Dehydroepiandrosterone (DHAS): <250 micro g/dl
Sex Binding Hormone Globulin: 18-114 nmol/l

Normal ranges- men

Total testosterone: 270-1100ng/dl
Free (Unbound) testosterone: 0.95-4.3 ng/dl

Appendix 2

Conference Participants

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