дифференцировки: критический взгляд на неразрешенные противоречия

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13 сентября 2020г.

Санкт-Петербург

Неопределенного вида гениталии у новорожденного являются, sine qua non*, парадигмой DSD

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* - sine qua non - без чего не обойтись

Consequences of the ESPE/LWPES guidelines for diagnosis and treatment of disorders of sex

Best Practice & Research Clinical Endocrinology & Metabolism

Key words: Intersex; Disorder of sex development (DSD); Consensus; Disclosure; Consent; Ethics.

> *Ambiguous genitalia of the newborn is, sine qua non, the paradigm of a disorder of sex development (DSD)*

that demands a multidisciplinary team approach to management. The problem is immediately apparent at birth, and what is conveyed to the family in the ensuing hours and days will have a long-lasting impact. Assignment to either...

В составе междисциплинарной команды

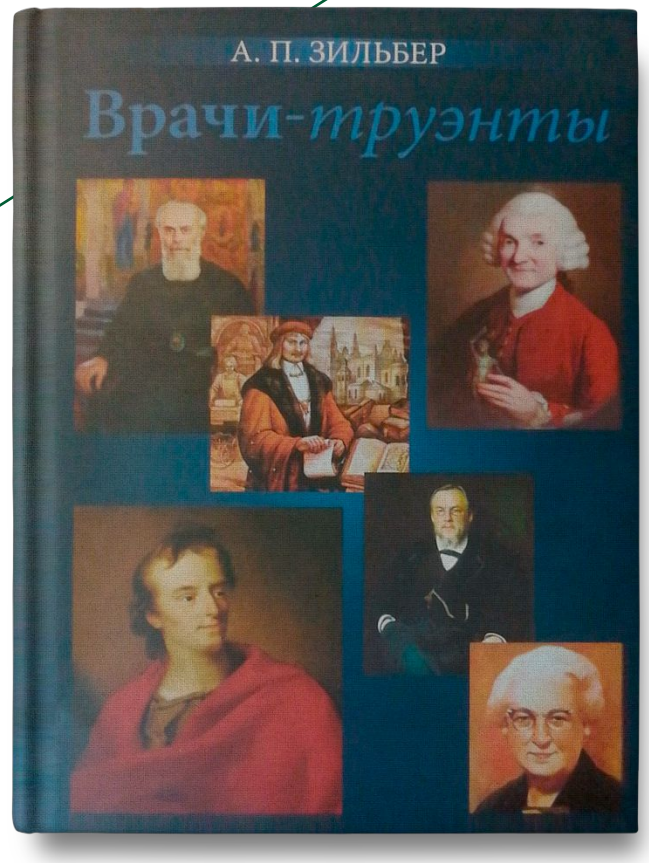
Мы должны сообща определять тактику лечения детей,

имеющих при рождении неопределенные (в отношении половой принадлежности) гениталии (или гениталии, вид которых допускает двоякое толкование) и нуждающихся, так или иначе, в хирургической коррекции

Для этого необходимо понимать друг друга

Терминология/номенклатура

Римский бог Терминус



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Pierre Mouriquand

Commentary to «Attitudes towards ‘disorders of sex development’ nomenclature among affected”

«In the beginning was the Word …» [1].

В начале было Слово… - Ин. 1:1

The authors should be congratulated for this excellent article [2] raising the essential issue of terminology to identify individuals born with atypical genitalia. The emotional impact of words describing these situations was evaluated by a cohort of patients and caregivers. There was clearly negative feedback from many patients, associations, and caregivers regarding the acronym DSD (disorders of sex development), which is uncomfortable, stigmatizing, and confusing for many in the current debate on management of these conditions.

The authors focused on the AIS group to evaluate the impact of terminology, which represents a limitation in their study, as the DSD issue covers many distinct groups of pathologies which may lead to different personal perceptions. As an example, some CAH (congenital adrenal hyperplasia) groups openly deagreed with their assimilation to DSD [3,4]. Language and culture also interfere with the interpretation of what DSD entails. The term sex does not have the same meaning in English/American and some other languages. Does it refer to the «individual inner identity” (brain or cortical sex), which is a subtle perception of one’s own identity; or does it refer to the «outer identity” (genital sex)? In that case, the

[1] Saint John the Evangelist. Prologue. John 1:1.

[2] Johnson EK, Rosoklija I, Fmlayson C. Chen D,
Yerkes EB, Madonna MB. et al. Attitudes towards «disorders of sex development» nomenclature among affected individuals. J Pediatr Urol 2017 May 8. pit: SI477-5131 (17)30183-3. http://dx. dot .org/10.1016/J. jpurol .2017.03.03 5 [Epub ahead of print).

[3] Lin-Su K, Lekarev 0, Poppas DP, Vogiatzi MG. Congenital adrenal hyperplasia patient perception of ‘disorders of sex development’ nomenclature. Int J Pediatr Endocrinol

Терминология в англоязычной литературе:

PEDIATRICS PERSPECTIVES

Bringing Back the Term “Intersex”

disorders
of sex development (DSD)

differences
of sex development

variations
of sex development

Терминология в отечественной литературе

нарушения/патология/особенности полового развития

нарушения/аномалии/особенности формирования пола

вариации половой дифференцировки

нарушения дифференцировки пола

неопределенная половая принадлежность

неопределенность пола и т.д.

При этом некоторые авторы, использующие термин «нарушения формирования пола», с одной стороны почему-то считают, что именно он и был предложен на конференции в Чикаго, а с другой –

К терминологии

«Нарушения полового развития» – прямой перевод «disorders of sex development (DSD)». При этом, строго говоря, не совсем ясно, о каком развитии идет речь: об эмбриональном развитии половых органов или о половом развитии ребенка – оба могут быть нарушены.

Наилучшим выбором может быть «нарушения половой дифференцировки» – «disorders of sex differentiation (DSD)»

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Julia Spencer
Barthold

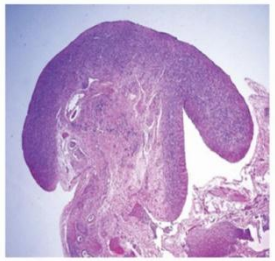
Disorders of Sex Differentiation: A Pediatric Urologist's Perspective of New Terminology and Recommendations

Purpose: In 2005 medical and lay experts convened (the Chicago Consensus), and reviewed and updated nomenclature and treatment recommendations in individuals with congenitally atypical gonadal, chromosomal or anatomical gender. This review summarizes, analyzes and considers the implications of these recommendations in pediatric urology practice.
Materials and Methods: Publications identified in a PubMed® search of 2000 to 2010 as well as relevant prior reports of new concepts and trends in the diagnosis of and treatment for intersex/ambiguous genitalia/disorders of sex differentiation, and responses to the Chicago Consensus were reviewed.
Results: In response to concerns regarding outdated, confusing and/or controversial terms, such as "intersex," "hermaphroditism" and "sex reversal," help more clearly determine appropriate management and prognosis for this heterogeneous group of disorders.

"the consensus statement recommended a new taxonomy based on the umbrella term, "disorders of sex differentiation."

Additional categorization based on sex chromosome complement was recommended but not clearly defined and variously interpreted. Routine use of multidisciplinary diagnostic and expert surgical teams, continuing psychosocial and psychosexual care, and full disclosure of alternatives relating to surgery type and timing were recommended. Early gender assignment was advocated but evidence-based guidance to support some aspects of care of affected individuals was insufficient. Pediatric urologists should remain abreast of new data refining the diagnoses and outcomes of disorders of sex differentiation, and ensure that their patients have access to multidisciplinary resources. Conclusions: Major changes in classification and expectations in the

Терминология в отечественной литературе



Вызывает вопросы:

Описание НПО, как имеющих

- интерсексуальное строение
- гермафродитное строение
- двойственное строение и т.д.

Представление о том, что патология уретры и влагалища у девочек с выраженной вирилизацией является **женской гипоспадией**

Разночтение в понимании, что такое **streak гонада** и как правильно произносится слово **streak [stri:k].**

“Streak gonad composed of ovarian stroma with no identifiable follicular structures”.

Классификация

Пример классификации Чикагского консенсуса:

> *While consideration of karyotype is useful br da ssifi cat bn, unnecessary reference to karyotype should be avoided; ideally, a system based on descriptive terms (for example, androgen insensitivity syndrome) should be used wherever possible.*

AMH, anti-mullerian hormone; CAIS, complete androgen insensitivity syndrome; DSD, disorders of sex development; MURCS, mullerian, renal, cervicothoracic somite abnormalities; PAIS, partbl androgen insensitivity syndrome; POR, cytochrome P450 oxidoredudase.

An example of a DSD classification

anomaly	46,XY DSD	46,XX DSD
Sex chromosome		
(A) 45 ,X (Turner syndrome and variatfs)	(A) Disorders of gonadal (tetficula-) development	(A) Disorders of gonadal (ovarian) d^elopment
	1. Complete gonadal dysgenesis (Swyer syndrome)	1. Ovotesticular DSD
(B) 47.XXY (Klinefelter synrfrome and variants)	2. Partial gonadal dysgenesis 3. Gonadal regression	2 Testicular DSD (eg, SRY+, dup SOX9) 3. Gonadal dysgenesis
	4. Ovofeslicular DSD	
(Q 45.X/46.XY (mixed go nodd dysgenesis, ovofeslicular DSD)	(B) Disorders in androgen synthesis or action 1. Androgen biosynthesis delect (eg, 17* hydroxysleroid dehydrogenase deficiency, 5* reductase deficiency, StAR mutations 2. Delect in androgen odbn (eg, CAIS, PAIS) 3. IH receptor defects (eg, Leyd'g cell hypoplasia, aplasia) 4. Disorders of AMH and AMH receptor (persistent mullerian duct syndrome)	(B) Androgen excess 1. Fetal (eg, 21-hydroxyfase deficiency, 11-hydraxylase deficiency) 2 Fetoplacental (aromatase deficiency, POR) 3. Maternd (luteoma, exogenous, etc)
(D) 46,XX/46,XY (chimenc, ovotesticular DSD)	(Q Other (eg. severe hypospadias, doocal extrophy)	IQOber (eg, doacd erfrophy, vaginal atresia, MURCS, other syndromes)

К терминологии

“В настоящее время основными категориями DSD являются 46,XX DSD, 46,XY DSD, sex chromosome DSD, ovotesticular DSD и 46,XX testicular DSD”

Sarah M. Lambert, MD*. Eric J.N. Vilain, MO.
PhDb,
Thomas F. Kalon, MO*–*

A Practical Approach to Ambiguous Genitalia in the Newborn Period

Keywords:
Ambiguous genitalia, Congenital adrenal hyperplasia, Disorders of sex development, Neonates

CHAPTER

The evaluation and management of a newborn with ambiguous genitalia must be undertaken with immediacy and great sensitivity. The pediatric urologist endocrinologist geneticist, and child psychiatrist or psychologist should work closely with the family in pursing a dual goal: to establish the correct diagnosis of the abnormality and. with input from the parents, determine gender based on the karyotype, endocrine function, and anatomy of the child. In this section the authors outline a practical approach to the neonate born with a disorder of sex development (DSD).

Nomenclature

Genital ambiguity in the neonate has been described for centuries and evidence for disorders of sexual differentiation exists from many ancient civilization’ The actual incidence of DSD is difficult to accurately determine because of the heterogeneity of the clinical presentation and the varied etiologies. Using birth registries, some authors have attempted to estimate the incidence of ambiguous genitalia at

The main categories include sex chromosome DSD. 46.XX DSD. and 46XY DSD. Some conditions can be placed into more than one category. Additionally, although tho majority of infants with 46XX DSD will be diagnosed with congenital adrenal hyperplasia (CAH). only approximately 50% of children with 46. XY DSD will have a definitive clinical diagnosis.”

Diagnosis

Chromosomal sex is established at fertilization and the undifferentiated gonads subsequently develop into either testes or ovaries. A child's phenotypic sex results from the differentiation of internal ducts and external genitalia under the influence of hormones and transcription factors. Any disturbance among these processes results in ambiguous genitalia or DSO

Currently, the main categories of DSO are 46.XX DSD. 46.XY DSD. sex chromosome DSD. ovotesticular DSD. and 46XX testicular DSD)

Вариант классификации

Best Practice & Research
Clinical Endocrinology &
Metabolism

Consequences of the ESPE/LWPES guidelines for diagnosis and treatment of disorders of sex

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Table 2. A proposed classification of causes of disorders of sex development (DSDs).

Sex chromosome DSD	46,XY DSD	46,XX DSD
A: 47,XXY (Klinefelter syndrome and variants) B: 45,X (Turner syndrome and variants) C: 45,X/46,XY (mixed gonadal dysgenesis) D: 46,XX/46,XY (chimerism)	A: Disorders of gonadal (testicular) development 1. Complete or partial gonadal dysgenesis (e.g. SRY, SOX9, SF1, WT1, DHH etc) 2. Ovotesticular DSD 3. Testis regression	A: Disorders of gonadal (ovarian) development 1. Gonadal dysgenesis 2. Ovotesticular DSD 3. Testicular DSD (e.g. SRYþ, dup SOX9, RSP01)
	B: Disorders in androgen synthesis or action 1. Disorders of androgen synthesis LH receptor mutations SmitheLemlieOpitz Syndrome Steroidogenic acute regulatory protein mutations Cholesterol side-chain cleavage(CYP11A1) 3b-hydoxysteroid dehydrogenase 2 (HSD3B2) 17a-hydroxylase/17,20-lyase (CYP17) P450 oxidoreductase (POR) 17b-hydoxysteroid dehydrogenase(HSD17B3) 5a-reductase 2 (SRD5A2) 2. Disorders of androgen action Androgen Insensitivity Syndrome Drugs and environmental modulators	B: Androgen excess 1. Fetal 3b-hydoxysteroid dehydrogenase 2 HSD3B2 21-hydroxylase (CYP21A2) P450 oxidoreductase (POR) 11b-hydoxylase (CYP11B1) Glucocorticoid receptor mutations 2. Fetoplacental Aromatase (CYP19) deficiency Oxidoreductase (POR) deficiency 3. Maternal Maternal virilizing tumours (e.g. luteomas) Androgenic drugs
C: 45.X/46.XY (mixed gonadal dysgenesis)	C: Other 1. Syndromic associations of male genital development (e.g. cloacal anomalies, Robinow, Aarskog, Hand-FootGenital, popliteal pterygium) 2. Persistent Mu¨llerian duct syndrome 3. Vanishing testis syndrome 4. Isolated hypospadias (CXorf6) 5. Congenital hypogonadotropic hypogonadism 6. Cryptorchidism (INSL3, GREAT)	C: Other 1. Syndromic associations (e.g. cloacal anomalies) 2. Mu¨llerian agenesis/hypoplasia e.g. MURCS) 3. Uterine abnormalities (e.g. MODY5) 4. Vaginal atresis (e.g. KcKusickeKaufman) 5. Labial adhesions

Классификация: расширенное толкование DSD

«DSD – это врожденные состояния, при которых хромосомный, гонадный или анатомический пол является истинно ненормальным»

Тем не менее проф.Hughes, участвовавший в принятии Чикагского Консенсусом определения DSD, далее писал, что спайкообразование между половыми губами у девочки является нарушением полового развития, поскольку ее мать обеспокоена тем, имеется ли у дочери нормальное влагалище!

Journal of Pediatric Urology (2010)
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Ieuan Hughes

How should we classify intersex disorders?

It is a tall order to expect 50 experts on a subject in medicine to reach unanimity when tasked with devising an alternative nomenclature and classification system for a set of conditions that manifest as intersex at birth or at puberty with somatic sex characteristics discordant with sex assignment. Yet, that was attempted in 2005 and realized as what has now become known as the Chicago Consensus on management of intersex disorders [1]. The task was approached using the strategy of consensus decision making, which involves reaching general agreement or an accord amongst a group of individuals. While it is acknowledged that some participants may express divergent views, they are nevertheless willing to accede to the ethos that the sum of the parts is more important than the individual components. This enables a concordat to be reached for which the group as a whole is responsible. When such a consensus document reaches the public domain, it is inevitable that experts in the subject area will exercise their right to dissent over certain elements. Such debate is to be welcomed, for which an opportunity has arisen in this issue of the Journal based on the paper by the Aaronsons [2].

> *even the common labial adhesion which can completely occlude the vaginal opening is not a 'trivial' matter for the mother who is concerned that her daughter does not have a normal vagina*

So what is the basis for the authors now proposing an alternative classification system for DSD and what are its merits? It is argued that the starting point should not be sex chromosomes as this is unreliable as a diagnosis. But knowing that a karyotype is XX in an infant with DSD is not a diagnosis, it merely steers the investigator towards one of the three subgroups. The first subgroup defined as a sex chromosome anomaly [will be readily identified by examples such as 47XXY, 45XO/46XY, 46XX/46XY and several other cases of aneuploidy that can arise. Where this karyotype-based approach fails,

Варианты классификации

«non-hormonal/non-chromosoma l DSD»

Journal of Pediatric Urology (2016)

Surgery in disorders of sex development (DSD) with a gender issue: If (why), when, and how?

Pierre D.E. Mouriquand, Daniela Brindusa Gorduza, Claire-Lise Gay, Heino F.L. Meyer-Bahlburg, Linda Baker, Laurence S. Baskin, Claire Bouvattier, Luis H. Braga Anthony C. Caldamonej, Lise Duranteau, Alaa El Ghoneimi, Terry W. Hensle, Piet Hoebeke, Martin Kaefer, Nicolas Kalfa, Thomas F. Kolon, Gianantonio Manzonis, Pierre-Yves Mure a,b, Agneta Nordenskjold, J.L. Pippi Salle, Dix Phillip Poppas, Philip G. Ransley A, Richard C. Rink, Romao Rodrigo, Leon Sann, Justine Schoberja, Hisham Sibai, Amy Wisniewski, Katja P. Wolffenbuttelad, Peter Lee and

Summary

Ten years after the consensus meeting on disorders of sex development (DSD), genital surgery continues to raise questions and criticisms concerning its indications, its technical aspects, timing and evaluation. This standpoint details each distinct situation and its possible management in

"non-hormonal/non chromosomal”

DSD) Questions are summarized for each DSD group with the support of literature and the feedback of several world experts.

Given the complexity and heterogeneity of presentation there is no consensus regarding the indications, the timing, the procedure nor the evaluation of outcome of DSD surgery. There are, however, some issues on which most experts would agree: 1) The need for identifying

5 main groups of DSD patients with atypical genitalia

«non-endocrine/malformative DSD»

Birth Defects Research (Part C) 00:000-000, 2014.

When Hormone Defects Cannot Explain It: Malformative Disorders of Sex Development

Romina P. Grinspon and Rodolfo A. Rey

Key words:

uterine and vaginal malformations; cloacal and bladder exstrophy, aphallia; hypospadias; penoscrotal transposition

The birth of a baby with malformations of the genitalia urges medical action. Even in cases where the condition is not life-threatening, the identification of the external genitalia as male or female is emotionally essential for the family, and genital malformations represent one of the most stressful situations around a newborn. The female or male configuration of the genitalia normally evolves during fetal life according to the genetic, gonadal, and hormonal sex. Disorders of sex development occur when mate hormone (androgens and anti-Müllerian hormone) secretion or action is insufficient in the 46,XY fetus or when there is an androgen excess in the 46,XX fetus. However, sex hormone defects during fetal development cannot explain all congenital malformations of the reproductive tract. This review is focused on those congenital conditions in which gonadal function and sex hormone target organ sensitivity are normal and, therefore, not responsible for the genital malformation. Furthermore, because the reproductive and urinary systems share many common pathways in embryo-fetal development, conditions associating urogenital malformations are discussed.

Классификация: имеющиеся проблемы

- Отсутствие единой общепризнанной классификации
- Изменчивость рубрик в предложенных, ряд из которых четко не определены и по-разному интерпретируются (complete/pure/partial/mixed gonadal dysgenesis)
- Сочетание старых и новых названий патологии
- Включение в одну группу больных, имеющих совершенно разные клинические характеристики, например, в группу 46,XX DSD девочек с ВДКН, имеющих вирилизацию НПО и нуждающихся в хирургической коррекции, и мальчиков (46,XX males) с нормальным фенотипом и не требующих никаких вмешательств

Все это создает серьезную путаницу, разобраться в которой бывает достаточно

Терминология и классификация

Таким образом, как представляется, назрела насущная **необходимость** унифицировать терминологию/номенклатуру и дать критическую оценку используемым классификациям.

Без общего языка трудно понимать друг друга

Рабочая классификация

Интерсекс	Нарушения половой дифференцировки (НПД = DSD)	Гонады
Женский псевдогермафродитизм	46,XX НПД	яичник + яичник
Мужской псевдогермафродитизм	46,XY НПД	яичко + яичко
Истинный гермафродитизм	Овотестикулярное НПД	яичко + яичник или овотестис
Смешанная дисгенезия гонад	Смешанная дисгенезия гонад	яичко + streak

Назначение пола

Pediatrics
Official journal of the American
academy of pediatrics

INVESTIGATION AND MANAGEMENT OF DSD

General Concepts of Care

Optimal clinical management of individuals with DSD21 should comprise the following: (1) gender assignment must be avoided before expert evaluation in newborns: <2) evaluation and long-term management must be performed at a center with an experienced multidisciplinary team; (3)

> *all individuals should receive a gender assignment*

(4) open communication with patients and families is essential, and participation in decision-making is encouraged: and (5) patient and family concerns should be respected and addressed in strict confidence.

> *Evidence supports the current recommendation to raise markedly virilized 46,XX infants with CAH as female.*

Approximately 60% of 5-a-reductase (5aRD2)-deficient patients assigned female in infancy and virilizing at puberty (and all assigned male) live as males.5 In 5aRD2 and possibly 170-hydroxysteroid dehydrogenase deficiencies, for which the diagnosis is made in infancy, the combination of a male gender identity in the majority and the potential for fertility (documented in 5aRD2 but unknown in 170-hydroxysteroid dehydrogenase deficiencies) should be discussed when providing evidence for gender assignment

Регистрация новорожденного без назначения пола

Возникающие при этом вопросы:

— Кто родился, мальчик или девочка?
— Как называть ребенка?
— Как его одевать?
— Как его воспитывать?
— Как на него будут реагировать другие дети?

и т.д., а также суждение о том, что «третьего» пола нет и быть не может, надеюсь, остаются для нас все еще актуальными

S F Ahmed
S Morrison
I A Hughes

Intersex and gender assignment: the third way?

to these usual goals of management by the child, family, and carers will vary from case to case.

> *A third gender is not a feasible option*

considering that in most cultures around the world, gender variants are not treated as equals and that the nations of die industrialized society are ill equipped to cope with this concept. Unless we decide that all individuals with complex genital anomalies live socially as "intersex people", this is not a simple solution

> *and may be considered as "sweeping the problem under die carpet"*

Назначение мужского пола при 46,XX CAH

Review of Outcome Information in 46,XX Patients with Congenital Adrenal Hyperplasia Assigned/Reared Male: What Does It Say about Gender Assignment?

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Should Male Gender Assignment be Considere Virilized Patient With 46,XX and Congenital A

Peter A. Lee,* Christopher P. Houk and Douglas A. Husmar

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Abbreviations and Acronyms

CAH = congenital adrenal hyperplasia

DSD = disorders of sex development

SCL-90 = Hopkins Symptom Checklist

Study received institutional review board approval.

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Purpose: We assess the outcome in 4 who were born with Prader 4 or 5 ge

Materials and Methods: After recei subject consent we reviewed the med 46,XX congenital adrenal hyperpla issue questionnaires.

Results: All subjects were assigned virilizing congenital adrenal hyperpla male gender identity with sexual orie always lived as male and 2 who we subsequently self-reassigned as mal partners, including 7 married 12 yea female partner included 1 priest, 1 divorced and self-reassigned as male, except the priest and the subject who

Special Feature
Approach to the patiean

Christopher P Houk, Peter A Lee

Approach to assigning gender in 46,XX Congenital Adrenal Hyperplasia with Male External Genitalia: Replacing Dogmatism with Pragmatism

The goal of sex assignment is to facilitate the best possible quality of life for the patient. Factors such as reproductive system development, sexual identity, sexual function, and fertility are important considerations in this regard. Although some DSD gender assignments are relatively straightforward, those with midstage genital ambiguity and unclear gonadal function represent a major challenge. A recent major change in DSD care has been to encourage a male assignment for 46,XY infants with ambiguous genitalia who have evidence of testicular function and in utero central nervous system androgen exposure. In contrast, assignment of virilized 46,XX DSD patients remains female when ovaries and internal organs are present, regardless of the extent of virilization of the external genitalia. In this paper, we

Хирургическое лечение: решение Консенсуса

Pediatrics
Official journal of the American
academy of pediatrics

Surgical Management

The surgeon has a responsibility to outline the surgical sequence and subsequent consequences from infancy to adulthood. Only surgeons with expertise in the care of children and specific training in the surgery of DSD should perform these procedures. Parents now seem to be less inclined to choose surgery for less severe ditoro- megaly.

> *Surgery should only be considered in cases of severe virilization (Prader III—V) and be performed in conjunction, when appropriate, with repair of the common urogenital sinus.*

Because orgasmic function and erectile sensation may be disturbed by clitoral surgery, the surgical procedure should be anatomically based to preserve erectile function and the innervation of the clitoris. Emphasis is on functional outcome rather than a strictly cosmetic appearance. It is generally felt that surgery that is performed for cosmetic reasons in the first year of life relieves parental distress and improves attachment between the child and the parents; the systematic evidence for this belief is lacking.

> *Currently, there is inadequate evidence in relation to establishment of functional anatomy to abandon the practice of early separation of the vagina and urethra.*

Хирургическое лечение: изменение при обновлении Консенсуса

При обновлении консенсусного решения в 2015 году 32 приглашенных эксперта не смогли прийти к соглашению относительно показаний и сроков оперативного лечения детей с DSD.

Hormone Research
in Paediatrics

Global Disorders of Sex Development Update since 2006: Perceptions, Approach and Care

Lee P.A., Nordenström A., Houk C.P., Ahmed S.F., Auchus R., Baratz A., Baratz Dalke K., Liao L.-M., Lin-Su K., Looijenga 3rd L.H.J., Mazur T., Meyer-Bahlburg H.F.L., Mouriquand P., Quigley C.A., Sandberg D.E., Vilain E., Witchel S., and the Global DSD Update Consortium

The goal of this update regarding the diagnosis and care of persons with disorders of sex development (DSDs) is to address changes in the clinical approach since the 2005 Consensus Conference, since knowledge and viewpoints change. An effort was made to include representatives from a broad perspective including support and advocacy groups. The goal of patient care is focused upon the best possible quality of life (QoL). The field of

Запрет хирургического лечения

В 2017 году ПАСЕ была принята резолюция о необходимости запрета любого лечения, в том числе хирургического, направленного на изменение половых характеристик, включая гонады, гениталии и внутренние половые органы, у детей с интерсексом без их добровольного и информированного согласия.

Parliamentary Assembly Assemblee parlamentaire

Julia Spencer Barthold

Promoting the human rights of and eliminating discrimination against intersex people

7.1. with regard to effectively protecting children's right to physical integrity and bodily autonomy and to empowering intersex people as regards these rights:

> *prohibit medically unnecessary sex-"normalizing" surgery*

7.1.1. … , sterilization and other treatments practiced on intersex children without their informed consent;

7.1.2. ensure that, except in cases where the life of the child is at immediate risk,

> *any treatment that seeks to alter the sex characteristics of the child, including their gonads, genitals or internal sex organs, is deferred until such time as the child is able to participate in the decision, based on the right to*

Хирургическое лечение: мнение ESPU

European Sociaty
for Paediatric Urology

Prof. Guy Bogaert

Open letter to the Council of Europe

Counseling parents and children with DSD in a patient- and family-centered multidisciplinary setting should be complete and unbiased, and based on available scientific and condition-related outcome information. We also encourage patients and parents to obtain information from other sources, especially from patient support societies. :

We have learned that a 'one size fits all' treatment does not exist for patients with DSD: treatment should be tailored to individual needs, taking into account all medical, psychological, social, and cultural considerations of the patient and its parents.

All treatment options, including the pros and cons of each choice, are discussed extensively and repeatedly to ensure a well-considered shared decision.

Advocating a ban on medico-surgical treatment contradicts the atmosphere of equality, openness and trust as is currently provided in the various patient- and family-centers for DSD care, and is actually a step backwards in evolution rather than forwards.

We call society to entrust the care of children with DSD to their well-informed, committed parents and dedicated professionals of a multidisciplinary center.

Хирургическое лечение: консенсус ESPU и SPU

Сложные медицинские проблемы должны оставаться в компетенции семьи и квалифицированной медицинской команды, а не законодателей

ESPU – SPU Consensus statement
2020

Management of Differences of Sex development (DSD)

Conclusion

Complex medical problems should remain in the purview of the family and the expert medical team and thus should not be legislated. Banning surgery for all patients with DSD conditions is equally as harmful to individualized care as demanding surgery for all patients with DSD conditions. Clearly, neither approach is correct.

Хирургическое лечение: конструктивный взгляд

Почему назначение женского пола и ранняя хирургическая коррекция у девочек с ВДКН вызывают столько вопросов, тогда как их нет при назначении мужского пола и проведении раннего оперативного лечения у мальчиков с гипоспадией?

И это при том, что обе группы больных очень похожи:

- в обеих хромосомный и гонадный пол однозначны, имеются лишь аномалии строения НПО, по поводу которых может быть проведена успешная хирургическая коррекция с достижением альных сексуальной и нкций
- в обеих группах гендерная дисфория является редкой, несмотря на то, что отдаленные результаты проведенных операций далеки от идеальных

frontiers in Pediatrics

Should CAH in Females Be Classified as DSD?

Ricardo Gonzalez and Barbara M. Ludwikowski

Чикагский консенсус 2005 г.

Поворотный пункт в оказании медицинской помощи детям с интерсексом

Конец старой эры

(эры Money)

Consensus Statement on Management of Iutersex Disorders Peter A. Lee. Christopher P. Honk. S. Faisal Ahmed, Ienan A. Hughes and in collaboration with the participants in the International Consensus Conference on Intersex organized by the Lawson Wilkins Pediatric Endocrine Society and the European Society for Paediatiic Endocrinology Pediatrics 2006

Management framework paradigms for disorder of sex development

ABSTRACT

Until 2005, questions regarding medical treatment and diagnostic information on Disorders of Sex Development (DSD) were not systematically discussed with both the patients and their families; however, the way these patients are currently treated have been changing with time. Interventional changes in the clinical-psychotherapeutic-surgical areas of DSD determine not only different medical recommendations but also help to place the patient and the family into the decisional process of therapy. We must consider

two paradigmatic periods

that have influenced and transformed the clinical management framework of patients with DSD:

The "Money era" (1955)

which emphasized the role of the gonads as the diagnostic criterion, having the environment as determinant of the sex identity; and

The Chicago Consensus (2005)

phase, in which the role of genetics and molecular biology was critical for an early identification, as well as in building a proper sex identity, emphasizing ethical questions and the "stigma culture" In addition, recent data have focused on the importance of interdisciplinary and statements on questions concerning Human Rights as key factors in treatment decision making. Despite each of these management models being able to determine specific directions and recommendations regarding the clinical handling of these patients, we verify that a composite of these several models is the clinical routine nowadays. In the present paper, we discuss these several paradigms, and pinpoint clinical differences and their unfolding regarding management of DSD patients and their families.

Тихая революция

“The revolution may have been quiet, but it has certainly been effective and achieved with the minimum morbidity”

“Революция, возможно, была тихой, но она, безусловно, была эффективной и осуществлена с минимальной болезненностью”

best practice
& research clinical endocrinology & metabolism

Ieuan A
A.Hughes

The quiet revolution

The approach to the management of disorders of sex development (DSD) has undergone major changes in recent years

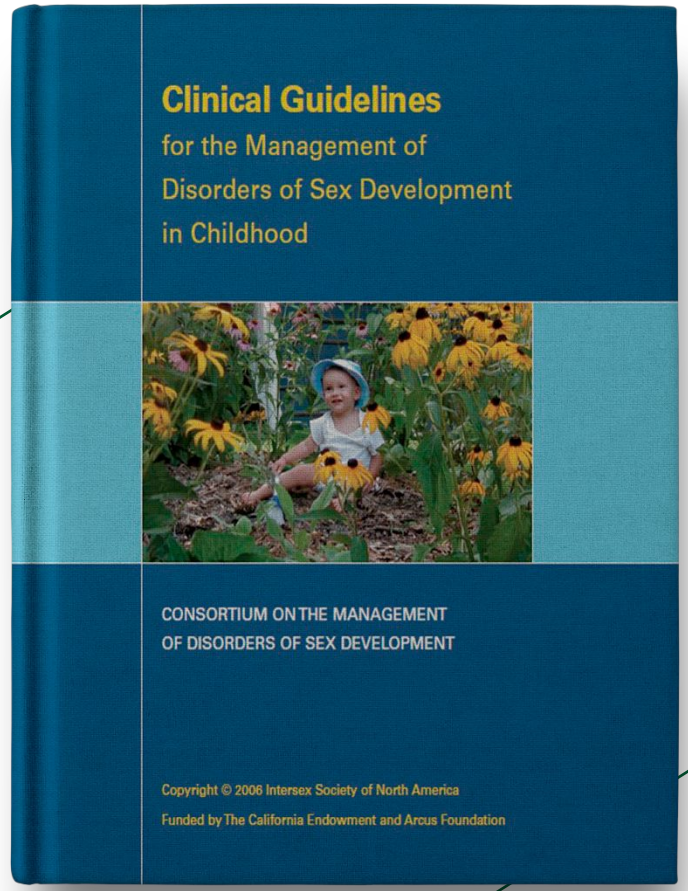
The catalyst has been a revised nomenclature, new classification

of the causes of DSD and a willingness for health professionals to work in a multi-disciplinary format. In a remarkably short length of time, these

revolutionary changes

are becoming accepted practice across a range of medical and scientific disciplines

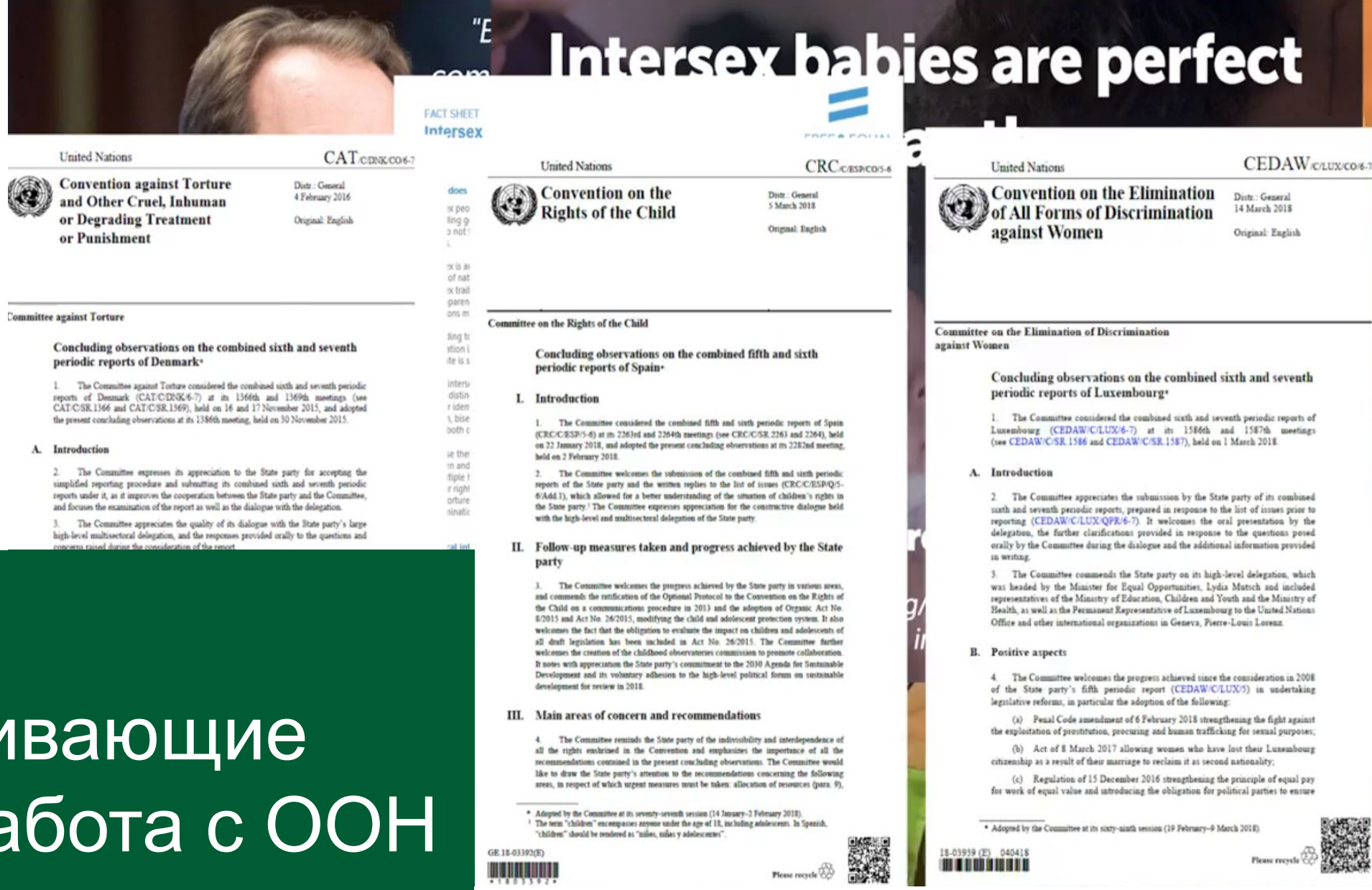
«Поддерживающие группы»: ISNA



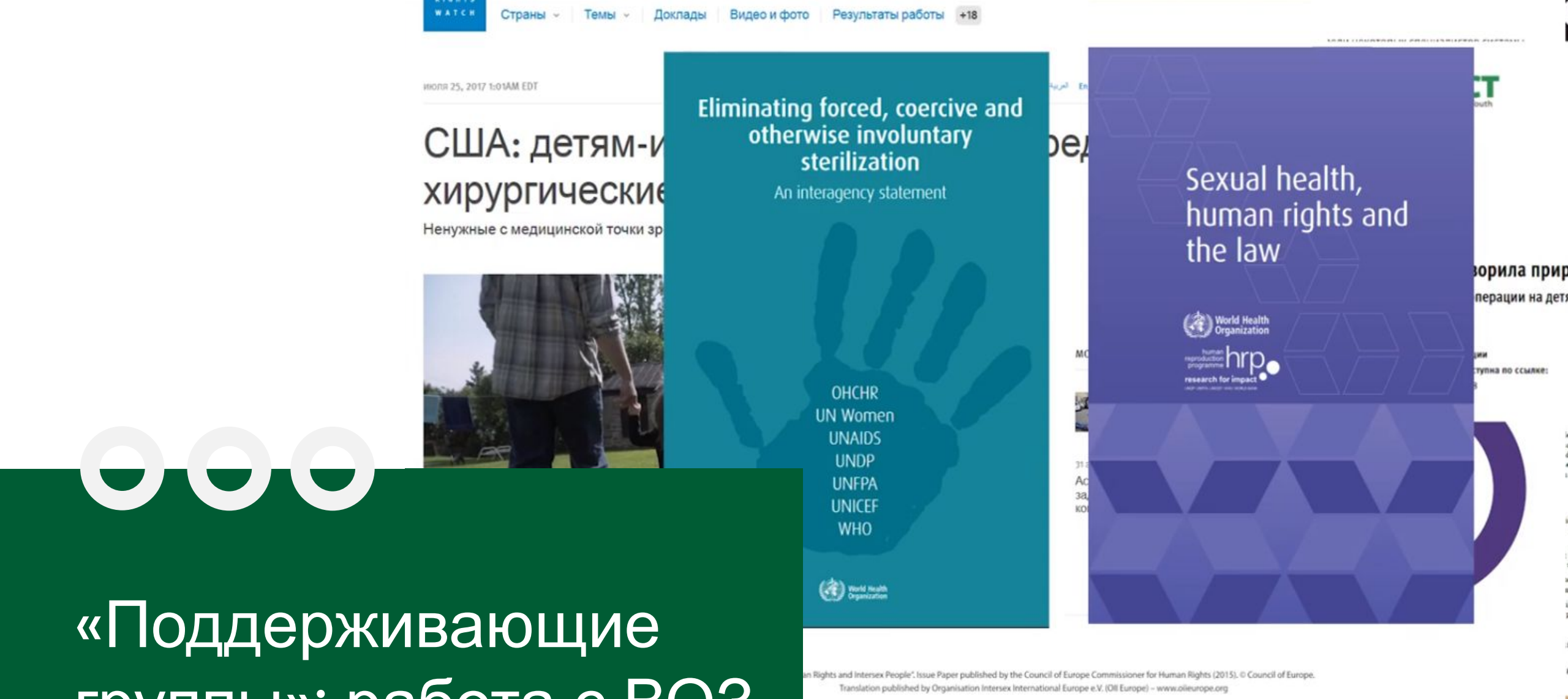
ISNA (1993) → Accord Alliance (2006)

peer support groups, patient advocacy groups, gender-right activist groups, intersex advocates…

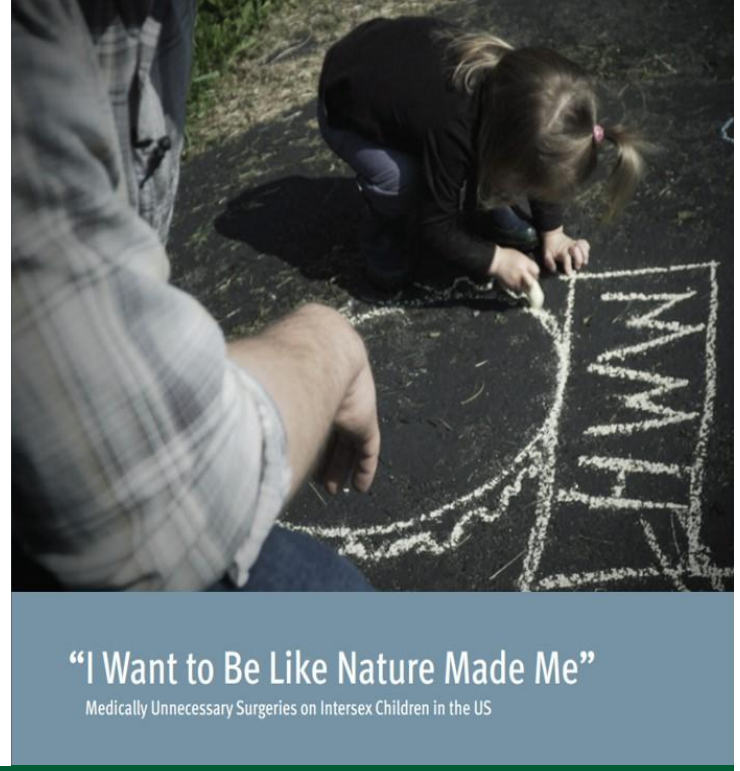
- При участии и под давлением ISNA были изменены терминология и классификация, подготовлены и изданы клинические рекомендации.
- Члены ISNA стали первыми настаивать на отсрочке назначения пола и оперативного лечения у детей до возраста, при котором те сами смогут определить свою половую (гендерную) идентичность и дать согласие на хирургическое вмешательство.

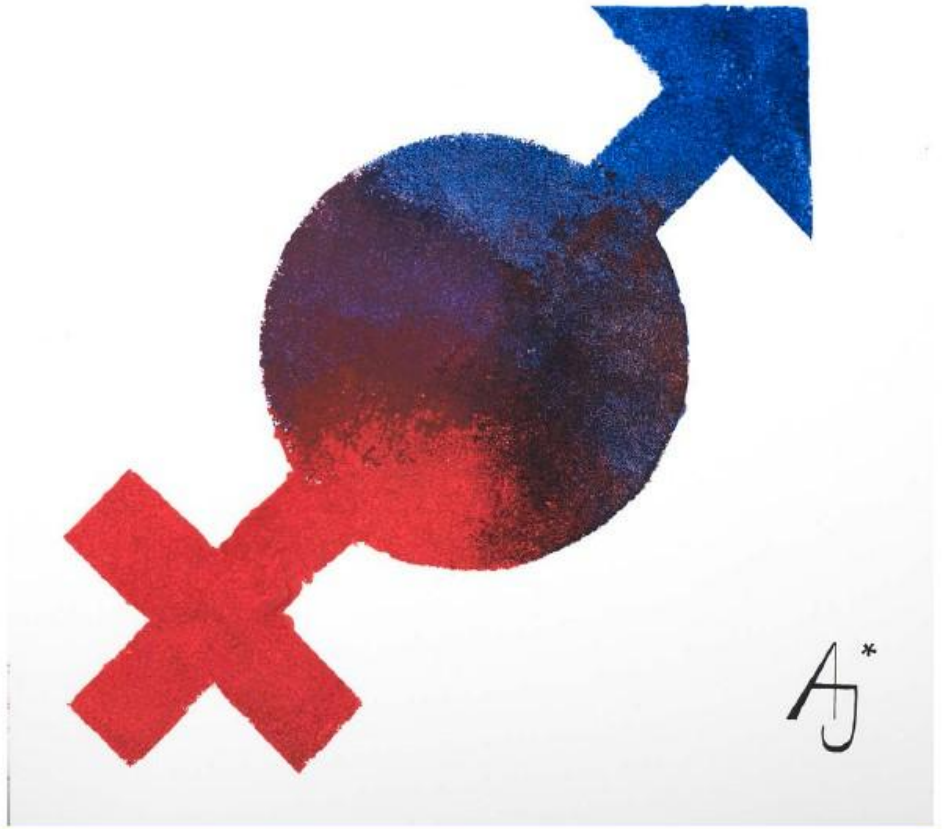


«Поддерживающие группы»: работа с ООН



«Поддерживающие группы»: работа с ВОЗ





«Поддерживающие группы»: работа с правозащитными организациями

«Поддерживающие группы»: взгляд на лечение

Хирургические вмешательства на гениталиях и гонадах у младенцев и детей с DSD характеризовались:

— как ненужные или косметические,
— как посягательства на физическую целостность (bodily integrity), на права ребенка, в том числе на «открытое будущее».

Феминизирующая генитопластика приравнивалась:

— к нанесению увечья ребенку (Female Genital Mutilation = Intersex Genital Mutilation),
— жестокому обращению с ним,
— и даже к пыткам.

Врачи-хирурги назывались

«Поддерживающие группы»: работа с ООН

Human Rights Council
Twenty-eighth session
Agenda item 3

Juan E. Méndez

Report of the Special Rapporteur on torture and other cruel, inhuman or degrading treatment or punishment

"*torture*"

Promotion and protection of all human rights, civil,
political, economic, social and cultural rights, including the right to development

Addendum

Observations on communications transmitted to Governments and replies received

«Поддерживающие группы»: мнение ESPU и SPU

Ведущие специалисты Европейского и Американского обществ детских урологов вынуждены были реагировать на слово «пытка» и разъяснять, что проводимые у таких больных операции, включая феминизирующие, не являются по сути своей косметическими и имеют вполне конкретные медицинские показания.

Journal
of Pediatric urology

P Mouriquand, A Caldamone, P Malone, J D Frank, P Hoebeke

The ESPU/SPU standpoint on the surgical management of Disorders of Sex Development (DSD)

DSD management and more specifically surgical manage-ment of DSD has been the

> *target for much criticism comingfrom various sources including a recent UN report ontorture (!) and a Swiss ethical committee.*

Specialistsinvolved in DSD management are primarily represented bypaediatric urologists as well as paediatric endocrinologistswho are aware of the dissatisfaction expressed by some DSDpatients who feel that the treatment they received, several(sometimes

«Поддерживающие группы»: отсутствие единой реакции медицинского сообщества

Кажется странным, но медицинское сообщество на Западе, по сути, не выступало категорически против принимаемых решений, несмотря на то, что речь шла об очень серьезных вещах:

— изменении бинарного принципа назначения пола с возможностью регистрации новорожденного без такового или с «третьим» полом (который может называться по-разному),

— ущемлении права родителей принимать решения в отношении лечения их ребенка,

— полном запрещении проведения хирургического лечения у детей без учета социальных, культурных, этнических и религиозных обстоятельств.

«Поддерживающие группы»: ARSI

Наверное, можно считать, что ничего страшного не происходит, и все образуется. Однако, как представляется, мы должны серьезно оценивать эту ситуацию, а не «sweeping the problem under the

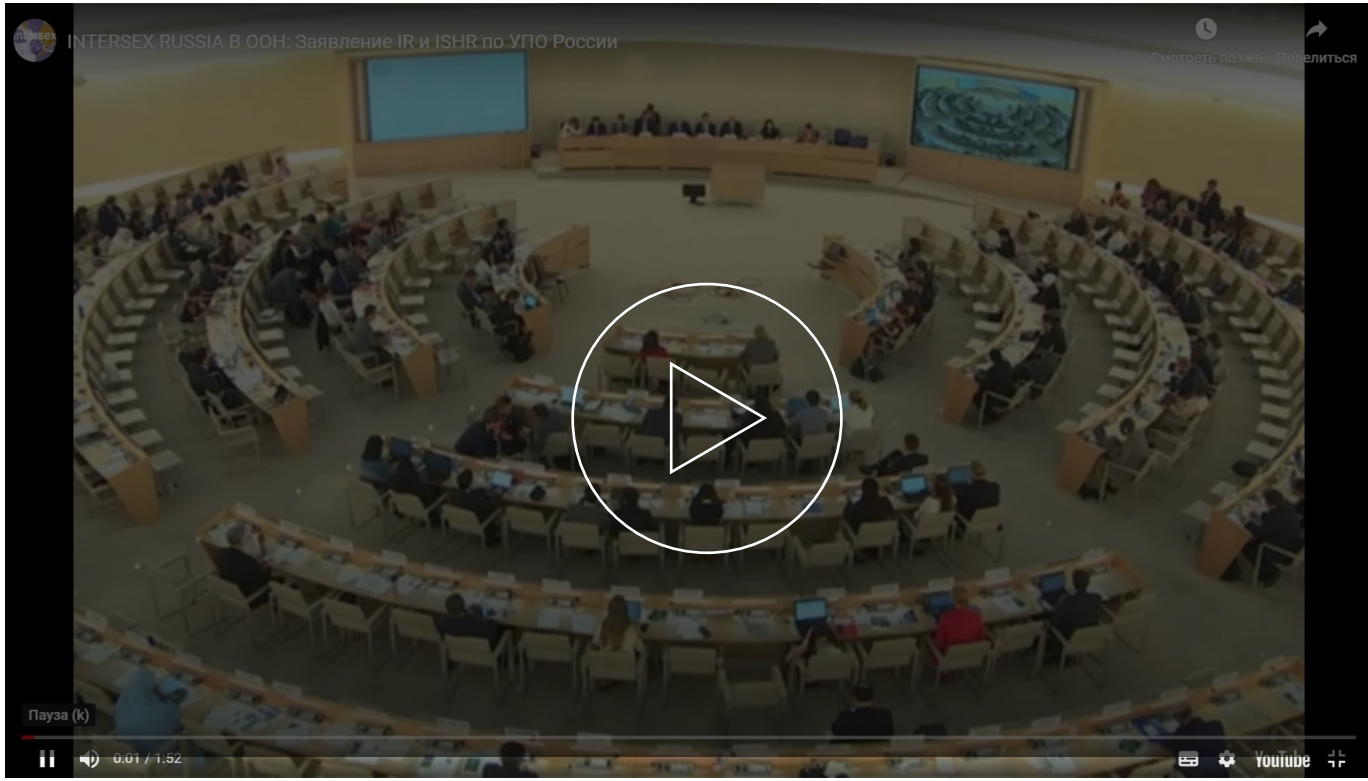
Association of Russian Speaking Intersex

Во-первых, думается, что многие наши специалисты, проповедовавшие этапное, т.е. позднее хирургическое лечение девочек с ВДКН, с радостью воспримут введение моратория.

Во-вторых, недавно мы получили письмо от одной уже **российской группы с такой преамбулой: «...естественное разнообразие не нуждается в коррекции...»** и просьбой «изменить подход к пациентам с **вариациями полового развития** с попыток излечить естественную особенность таких людей на поддержку и сопровождение при необходимости»(!)

Поддерживающие группы»: Intersex Russia

В-третьих, теперь у нас в стране появилась и вторая такая группа, представители которой выступили в ООН с заявлением о несоблюдении прав интерсекс детей в России.



«Поддерживающие группы»: работа в России

Выражаем благодарность сообществу Intersex Russia

ФГБОУ ВО «Санкт-Петербургский государственный педиатрический медицинский университет» МЗ РФ

А.Н. Тайц, К.Е. Белозеров, А.В. Гуслистова, К.А. Омельчук

ТАКТИКА ВЕДЕНИЯ СИНДРОМА АНДРОГЕННОЙ РЕЗИСТЕНТНОСТИ В РОССИЙСКОЙ ФЕДЕРАЦИИ И СОВРЕМЕННЫЙ ВЗГЛЯД НА ПРОБЛЕМЫ НАРУШЕНИЯ ДИФФЕРЕНЦИРОВКИ ПОЛА

> *Дети, имеющие нарушения формирования пола, часто подвергаются хирургическим и иным процедурам без наличия медицинских показаний и в возрасте, когда они не могут принять самостоятельного решения, а также подвергаются стигматизации и дискриминации со стороны общества. ООН призывает к соблюдения прав интерсекс-людей*

Надеюсь, что мрак рассеется, и
врачебные вопросы вновь начнут
решать специалисты, а не законодатели
под нажимом активистов

Благодарю
за внимание