

## What does it mean to be intersex?

"Intersex" is a general term used for a variety of conditions in which a person is born with a reproductive or sexual anatomy that doesn't seem to fit the typical definitions of female or male. For example, a person might be born appearing to be female on the outside, but having mostly male-typical anatomy on the inside. Or a person may be born with genitals that seem to be in-between the usual male and female types--for example, a girl may be born with a noticeably large clitoris, or lacking a vaginal opening, or a boy may be born with a notably small penis, or with a scrotum that is divided so that it has formed more like labia. Or a person may be born with mosaic genetics, so that some of her cells have XX chromosomes and some of them have XY. Though we speak of intersex as an inborn condition, intersex anatomy doesn't always show up at birth. Sometimes a person isn't found to have intersex anatomy until she or he reaches the age of puberty, or finds himself an infertile adult, or dies of old age and is autopsied. Some people live and die with intersex anatomy without anyone (including themselves) ever knowing. Which variations of sexual anatomy count as intersex? In practice, different people have different answers to that question. That's not surprising, because intersex isn't a discreet or natural category. What does this mean?

Intersex is a socially constructed category that reflects real biological variation. To better explain this, we can liken the sex spectrum to the colour spectrum. There's no question that in nature there are different wavelengths that translate into colours most of us see as red, blue, orange, yellow. But the decision to distinguish, say, between orange and red-orange is made only when we need it--like when we're asking for a particular paint colour. Sometimes social necessity leads us to make colour distinctions that otherwise would seem incorrect or irrational, as, for instance, when we call certain people "black" or "white" when they're not especially black or white as we would otherwise use the terms. In the same way, nature presents us with sex anatomy spectrums. Breasts, penises, clitorises, scrotums, labia, gonads--all of these vary in size and shape and morphology.

So-called "sex" chromosomes can vary quite a bit, too. But in human cultures, sex categories get simplified into male, female, and sometimes intersex, in order to simplify social interactions, express what we know and feel, and maintain order. So nature doesn't decide where the category of "male" ends and the category of "intersex" begins, or where the category of "intersex" ends and the category of "female" begins. "Humans decide". Humans (today, typically doctors) decide how small a penis has to be, or how unusual a combination of parts has to be, before it counts as intersex. Humans decide whether a person with XXY chromosomes or XY chromosomes and androgen insensitivity will count as intersex. In our work, we find that doctors' opinions about what should count as "intersex" vary substantially. Some think you have to have "ambiguous genitalia" to count as intersex, even if your inside is mostly of one sex and your outside is mostly of another. Some think your brain has to be exposed to an unusual mix of hormones prenatally to count as intersex--so that even if you're born with atypical genitalia, you're not intersex unless your brain experienced atypical development. And some think you have to have both ovarian and testicular tissue to count as intersex. Rather than trying to play a semantic game that never ends, we at ISNA take a pragmatic approach to the question of who counts as intersex. We work to build a world free of shame, secrecy, and unwanted genital surgeries for anyone born with what someone believes to be non-standard sexual anatomy. By the way, because some forms of intersex signal underlying metabolic concerns, a person who thinks she or he might be intersex should seek a diagnosis and find out if she or he needs professional

healthcare. Check our **contacts page** to get in touch with us (Working It Out) or the Sexual Health Branch for further diagnostic information.

### How common is intersex?

To answer this question in an uncontroversial way, you'd have to first get everyone to agree on "what counts as intersex" --and also to agree on what should count as strictly male or strictly female. That's hard to do. How small does a penis have to be before it counts as intersex? Do you count "sex chromosome" anomalies as intersex if there's no apparent external sexual ambiguity? (Alice Dreger explores this question in greater depth in her book "Hermaphrodites and the Medical Invention of Sex", 1998, Cambridge: Harvard University Press.)

Here's what we do know: If you ask experts at medical centres how often a child is born so noticeably atypical in terms of genitalia that a specialist in sex differentiation is called in, the number comes out to about 1 in 1500 to 1 in 2000 births. But a lot more people than that are born with subtler forms of sex anatomy variations, some of which won't show up until later in life. Below we provide a summary of statistics drawn from an article by Brown University researcher Anne Fausto-Sterling (Blackless, Melanie, Anthony Charuvastra, Amanda Derryck, Anne Fausto-Sterling, Karl Lauzanne, and Ellen Lee. 2000. [How sexually dimorphic are we? Review and synthesis.](#) *American Journal of Human Biology* 12:151-166. The basis for that article was an extensive review of the medical literature from 1955 to 1998 aimed at producing numeric estimates for the frequency of sex variations. Note that the frequency of some of these conditions, such as congenital adrenal hyperplasia, differs for different populations. These statistics are approximations.

Not XX and not XY	one in 1,666 births
Klinefelter (XXY)	one in 1,000 births
Androgen insensitivity syndrome	one in 13,000 births
Partial androgen insensitivity syndrome	one in 130,000 births
Classical congenital adrenal hyperplasia	one in 13,000 births
Late onset adrenal hyperplasia	one in 66 individuals
Vaginal agenesis	one in 6,000 births
Ovotestes	one in 83,000 births
Idiopathic (no discernable medical cause)	one in 110,000 births
Iatrogenic (caused by medical treatment, for instance progestin administered to pregnant mother)	no estimate
5 alpha reductase deficiency	no estimate
Mixed gonadal dysgenesis	no estimate
Complete gonadal dysgenesis	one in 150,000 births
Hypospadias (urethral opening in perineum or along penile shaft)	one in 2,000 births
Hypospadias (urethral opening between corona and tip of glans penis)	one in 770 births
Total number of people whose bodies differ from standard male or female	one in 100 births
Total number of people receiving surgery to "normalize" genital appearance	one or two in 1,000 births

### How do I know if I have an intersex condition?

ISNA is working to create a world free of shame, secrecy, and unwanted sexual surgeries for children born with anatomy that someone decided is not standard male or female. This is different from, for example, having a feeling that your

identity is different from most women (or men). People with intersex conditions generally don't have to search for evidence that they are intersexed; the evidence is in their own bodies. For instance, women who do not have ovaries, men who don't have testes, women who have no clitoris or inner labia, people who remember multiple genital surgeries during childhood and scars in their genital area and abdomen, people who have ambiguous genitalia. Sometimes people tell us that they have fairly typical genitals, but they think that they must have been born intersex and subjected to a sex change as an infant. Surgeons, even today, cannot create "normal" looking genitals, and surgery was much poorer decades ago. Thus, if you have genitals that look like most women (or men), then you were surely born with these genitals.

### **Intersex conditions**

The following is a list of conditions of sex development that sometimes involve "intersex anatomy"

#### **Androgen Insensitivity Syndrome (AIS)**

Androgen Insensitivity Syndrome, or AIS, is a genetic condition, inherited (except for occasional spontaneous mutations), occurring in approximately 1 in 20,000 individuals. In an individual with complete AIS, the body's cells are unable to respond to androgen, or "male" hormones. ("Male" hormones is an unfortunate term, since these hormones are ordinarily present and active in both males and females.) Some individuals have partial androgen insensitivity. In an individual with complete AIS and karyotype 46 XY, testes develop during gestation. The foetal testes produce mullerian inhibiting hormone (MIH) and testosterone. As in typical male foetuses, the MIH causes the foetal mullerian ducts to regress, so the foetus lacks uterus, fallopian tubes, and cervix plus upper part of vagina. However, because cells fail to respond to testosterone, the genitals differentiate in the female, rather than the male pattern, and Wolffian structures (epididymis, vas deferens, and seminal vesicles) are absent. The newborn AIS infant has genitals of normal female appearance, undescended or partially descended testes, and usually a short vagina with no cervix. Occasionally the vagina is nearly absent. AIS individuals are clearly women. At puberty, the testes are stimulated by the pituitary gland, and produce testosterone. Because testosterone is chemically very similar to oestrogen, some of the testosterone converts back to oestrogen ("aromatizes") in the bloodstream. This oestrogen produces breast growth, though it may be late. Women with AIS do not menstruate, and are not fertile. Because the development of pubic and underarm hair, in women as well as in men, depends upon testosterone, most AIS women have no pubic or underarm hair, but some have sparse hair. When an AIS girl is diagnosed during infancy, physicians often perform surgery to remove her undescended testes. Although removal of testes is advisable, because of the risk of cancer, ISNA advocates that surgery be offered later, when the girl can choose for herself. Testicular cancer is rare before puberty.

Vaginoplasty surgery is frequently performed on AIS infants or girls to increase the size of the vagina, so that she can engage in penetrative intercourse with a partner with an average size penis. Vaginoplasty surgery is problematic, with many failures. ISNA advocates against vaginal surgery on infants. Such surgery should be offered to, not imposed on, the pubertal girl, and she should have an opportunity to speak with adult AIS women about their sexual experience and about surgery in order to make a fully informed decision. Not all AIS women will choose surgery. Some women have successfully increased the depth of their vagina with a program of regular pressure dilation, using aids designed for that purpose. Contact the "AIS

Support Group": <http://www.medhelp.org/ais/> . Physicians and parents have been most reluctant to be honest with AIS girls and women about their condition, and this secrecy and stigma has unnecessarily increased the emotional burden of being different. Because AIS is a genetic defect located on the X chromosome, it runs in families. Except for spontaneous mutations, the mother of an AIS individual is a carrier, and her XY children have a 1/2 chance of having AIS. Her XX children have a 1/2 chance of carrying the AIS gene. Most AIS women should be able to locate other AIS women among siblings or maternal relatives.

\*Is there a test to find out if you have AIS?\* The answer depends upon exactly what you are looking for--diagnostic information, or carrier status. If were born with female genitals and testes, and have very sparse or absent pubic hair, you most likely have complete AIS. If you were born with ambiguous genitals and testes, there are a number of possible aetiologies, including partial AIS. Testing for partial AIS is more problematic than the complete form. Hormonal tests in a newborn with 46 XY karyotype and ambiguous genitals will show normal to elevated testosterone and LH, and a normal ratio of testosterone to DHT. A family history of ambiguous genitals in maternal relatives suggests partial androgen insensitivity. If you are wondering if you are a carrier, or if you know that you are a carrier and are wondering about the status of your foetus, genetic testing is possible. AIS has been diagnosed as early as 9-12 weeks gestation by chorionic villus sampling (sampling tissue from the foetal side of the placenta). By the 16th week it can be detected by ultrasound and amniocentesis. However, prenatal diagnosis is not indicated unless there is a family history of AIS. See the following for details of testing. Hodgins M. B., Duke E. M., Ring D.: Carrier detection in the testicular feminization syndrome: deficient 5 alpha-dihydrotestosterone binding in cultured skin fibroblasts from the mothers of patients with complete androgen insensitivity. *J. Med. Genet.* Jun 1984, 21, (3), p178-81. Batch J. A., Davies H. R., Evans B. A. J., Hughes I. A., Patterson M. N.: Phenotypic variation and detection of carrier status in the partial androgen insensitivity syndrome. *Arch. Dis. Childh.* 1993; 68: 453-457.

### **Aphallia**

Aphallia refers to being born without a penis, in a patient with otherwise typical male anatomy.

### **Clitoromegaly (large clitoris)**

Clitoromegaly means that the clitoris is larger than expected. This is a description, rather than a diagnosis. The most common cause of clitoromegaly is probably "CAH", see below.

### **Congenital Adrenal Hyperplasia (CAH)**

Congenital Adrenal Hyperplasia (CAH) is the most prevalent cause of intersex among people with XX chromosomes. About 1 in 10,000 to 18,000 children are born with congenital adrenal hyperplasia, but it does not cause intersex in those with XY chromosomes, so the prevalence of CAH-related intersex is about 1 in 20,000 to 1 in 36,000. \*Among the many causes of intersex, only CAH represents a real medical emergency in the newborn period.\* Click here to read about the "medical risks of CAH" both at birth and later in life. CAH occurs when there is a broken genetic "recipe" for making cortisone in the adrenal glands (the glands on top of the kidneys that make various hormones and add them to the blood stream). Because the recipe is broken, the adrenal glands, while trying to make cortisone, may make an unusually high level of other hormones that are "virilizing". That is, they can

make XX embryos have larger than average clitorises, or even a clitoris that looks rather like a penis, or labia that look like a scrotum. To be more specific, intersex occurs in relation to CAH when an anomaly of the adrenal function causes the synthesis and excretion of an androgen precursor, initiating virilization of a XX person in-utero. Because the virilization originates metabolically, masculinizing effects continue after birth. So CAH can cause the person to develop masculine-typical characteristics like dense body hair, a receding hairline, deep voice, prominent muscles, etc. Sex phenotype varies along the full continuum, with the possible added complication of metabolic problems which upset serum sodium balance. The metabolic effects of CAH can be counteracted with cortisone. The long term use of cortisone itself produces significant dependence and other side effects, all of which need to be explained honestly and openly. CAH also occurs in XY individuals, and requires medical attention as it has significant impacts on health, but it does not cause ambiguous genitalia. CAH doesn't cause intersex in XY individuals because their testes already produce so much testosterone that the added virilizing hormones don't really make a difference. However, untreated CAH can cause boys to have their puberty earlier than other boys. This can be a problem because it will stop them from growing taller (so they end up as very short adults) and because it can cause them to be very sexual well before other children their age are having such strong sexual thoughts and desires.

## **Hypospadias**

Hypospadias refers to a urethral meatus ("pee-hole") which is located along the underside, rather than at the tip of the penis. In some hypospadias, the meatus may be located on the underside of the penis, in the glans. In more pronounced hypospadias, the urethra may be open from mid-shaft out to the glans, or the urethra may even be entirely absent, with the urine exiting the bladder behind the penis. Hypospadias is extremely common. There are now many online groups where men and boys with hypospadias, their families, and parents of boys with hypospadias can talk things over. Click here to see our **Contacts** page.

## **I have a line along the underside of my penis**

Sometimes men write to ISNA, saying that they have a line down the underside of their penis, all the way to the anus, and they wonder if this is evidence of some sort of intersex condition. That line is called the "penile raphe." From conception until 7 weeks, foetuses have no sex difference--the genitals look fairly female. There are two sets of swellings ("urogenital swellings"), open in the middle, and a protuberance toward the front ("phallus"). After 7 weeks, for boys with typical development, the foetal testes produce testosterone, and the testosterone causes the urogenital swellings to swell, come together in the middle, and fuse, forming the scrotum and the underside of the penis. The line down the middle, called a "raphe", is just a reminder of how all humans start out with a common female genital anatomy until 7 weeks after conception. The "Toronto Hospital for Sick Children's Child Physiology website":

<http://www.sickkids.ca/childphysiology/cpwp/Genital/genitaldevelopment.htm>

has a very clear animation showing how foetus's sexual anatomy develops. If you click "genitals" on the left, then "genital formation" on the left, you'll find an animation that shows how boy genitals and girl genitals start out looking the same (from conception to week 7), then develop along different lines under the influence of hormones. If the foetus has an unusual level of certain hormones, or an unusually high or low ability to respond to them, then intersex appearance can result.

## **Klinefelter Syndrome**

Most men inherit a single X chromosome from their mother, and a single Y chromosome from their father. Men with Klinefelter syndrome inherit an extra X chromosome from either father or mother; their karyotype is 47 XXY. Klinefelter is quite common, occurring in 1/500 to 1/1,000 male births. The testes are small (about half typical size) and quite firm. After puberty, the ejaculate contains no sperm. Other effects of Klinefelter are quite variable. Boys with Klinefelter are usually born with male genitals that look like other boys. But at puberty, they may not virilize very strongly--they may not develop much body hair, or they may experience breast development. If the boy \*wishes\* to virilize, testosterone (either through injections or via patches) can help him to do so. Although most boys with Klinefelter Syndrome grow up to live as men, some do develop atypical gender identities, and some do develop female gender identities.

## **Micropenis**

\*Micropenis\* is a term used by doctors for when a person has a penis that is very small and is completely differentiated (developed like a typical penis) with the urethral meatus ("pee-hole") at the tip. The criteria for clinical diagnosis include that a person has: a 46,XY karyotype (typical male karyotype); testes that are either descended or undescended; an urethral meatus ("pee-hole") at the tip of the glans penis (that is, no hypospadias), and a stretched penis length at or below 2.5 standard deviations for age and stage of development.

## **Mosaicism involving "sex" chromosomes**

First, some background: A karyotype is a picture of the chromosomes in a cell. A karyotype is used to see what kinds of chromosomes a person has. It is created by taking a blood or tissue sample from a person, and then staining the chromosomes with dye and photographing them through a microscope. The photograph is then cut up and rearranged so that the chromosomes are lined up into corresponding pairs. The result is usually reported as the number and type of a person's chromosomes, such as 45,X (the individual has 22 pairs of matched chromosomes and one X chromosome, also known as "Turner Syndrome":

<http://www.isna.org/faq/conditions/turner>); 46,XX (the individual has 22 pairs of matched chromosomes and two X chromosomes); 46,XY (the individual has 22 pairs of matched chromosomes, one X chromosome and one Y chromosome); 47, XXY. A person is said to have a "mosaic karyotype" when he or she has one kind of karyotype in some of his or her cells, and a different karyotype in other cells. An example is when a person is said to have a 45,X/46,XX karyotype; that means he or she has 46,X in some cells, and 46,XX in other cells. Mosaicism happens because sometimes cells divide incorrectly early in the life of an embryo. For instance a woman with Mosaic Turner Syndrome may have some cells that are XO (typical Turner Syndrome karyotype) and some cells that are XX (typical female karyotype). Mosaicism also occurs in milder forms of "Klinefelter Syndrome":

<http://www.isna.org/faq/conditions/klinefelter>, called 46/47 XY/XXY mosaic. In this case, the XY cells would have 46 chromosomes (a typical number of chromosomes) and the XXY cells would have 47 chromosomes.

## **MRKH (Mullerian agenesis; vaginal agenesis; congenital absence of vagina)**

Ovaries present with uterus absent, misshapen, or small; associated with kidney and spine anomalies in a minority of individuals. For more information, please check out "mrkh.org": <http://www.mrkh.org>, founded and run by former ISNA board member "Esther Morris Leidolf". A "Parent/Guardian Guide on MRKH":

[http://www.youngwomenshealth.org/mrkh\\_parent.html](http://www.youngwomenshealth.org/mrkh_parent.html) is also available from the Boston Children's Hospital.

### **Ovo-testes (formerly called "true hermaphroditism")**

\*Ovotestes\* are gonads (sex glands) containing both ovarian and testicular tissue. These are sometimes present in place of one or both ovaries or testes. In other words, a person might be born with two ovotestes, or a person might be born with one ovary and one ovotestes, or a person might be born with some other combination. The fact that a person has ovotestes won't tell you what his or her genitals looked like when he or she was born. Some people with ovotestes look fairly typically female, some fairly typically male, and some look fairly in-between in terms of genital development. Testicular tissue in ovotestes involves an increased risk of gonadal cancer. For this reason, people with ovotestes need to have the testicular portion removed or to at least be very carefully monitored by doctors.

### **Partial Androgen Insensitivity Syndrome (PAIS)**

The extent of androgen insensitivity in 46 XY individuals is quite variable, even in a single family. Partial androgen insensitivity typically results in "ambiguous genitalia." The clitoris is large or, alternatively, the penis is small and hypospadiac (these are two ways of labelling the same anatomical structure). Partial androgen insensitivity may be quite common, and has been suggested as the cause of infertility in many men whose genitals are of typically male appearance. Individuals with ambiguous genitalia have typically been subjected to "corrective" surgery during infancy. Based on our own painful experiences, ISNA believes that such cosmetic surgery of the genitals is harmful and unethical. Surgery is justified only when it is necessary for the health and well-being of the child. Surgery which is intended to make the genitals appear more male or more female should be offered, but not imposed, only when the child is old enough to make an informed decision for her/himself. \*Is there a test to find out if you have androgen insensitivity?\* The answer depends upon exactly what you are looking for-- diagnostic information, or carrier status. If were born with female genitalia and testes, and have very sparse or absent pubic hair, you most likely have complete AIS. If you were born with ambiguous genitalia and testes, there are a number of possible aetiologies, including partial AIS. Testing for partial AIS is more problematic than the complete form. Hormonal tests in a newborn with 46 XY karyotype and ambiguous genitalia will show normal to elevated testosterone and LH, and a normal ratio of testosterone to DHT. A family history of ambiguous genitalia in maternal relatives suggests partial androgen insensitivity. If you are wondering if you are a carrier, or if you know that you are a carrier and are wondering about the status of your foetus, genetic testing is possible. AIS has been diagnosed as early as 9-12 weeks gestation by chorionic villus sampling (sampling tissue from the foetal side of the placenta). By the 16th week it can be detected by ultrasound and amniocentesis. However, prenatal diagnosis is not indicated unless there is a family history of AIS. See the following for details of testing. Hodgins M. B., Duke E. M., Ring D.: Carrier detection in the testicular feminization syndrome: deficient 5 alpha-dihydrotestosterone binding in cultured skin fibroblasts from the mothers of patients with complete androgen insensitivity. *J. Med. Genet.* Jun 1984, 21, (3), p178-81. Batch J. A., Davies H. R., Evans B. A. J., Hughes I. A., Patterson M. N.: Phenotypic variation and detection of carrier status in the partial androgen insensitivity syndrome. *Arch. Dis. Childh.* 1993; 68: 453-457.

## **Progestin Induced Virilization**

Caused by prenatal exposure to exogenous androgens, most commonly progestin. Progestin is a drug which was administered to prevent miscarriage in the 50's and 60's and it is converted to an androgen (virilizing hormone) by the prenatal XX persons metabolism. If the timing is right, the genitals are virilized with effects ranging from enlarged clitoris to the development of a complete phallus and the fusing of the labia. In all cases ovaries and uterus or uterine tract are present, though in extreme cases of virilization there is no vagina or cervix, the uterine tract being connected to the upper portion of the urethra internally. The virilization only occurs prenatally and the endocrinological functionality is unchanged, ie. feminising puberty occurs due to normally functioning ovaries. In other words, XX people affected in-utero by virilizing hormones can be born into a continuum of sex phenotype which ranges from "female with larger clitoris" to "male with no testes". It is noteworthy that the use of progestin is not effective in the prevention of miscarriage. Progestin androgenized children are subjected to the same surgically enforced standards of cosmetic genital normalcy as other intersexed children... meaning that clitoridectomy and possibly more extensive procedures are often performed early in life, most often with the effect of loss of erotic sensation and ensuing psychological trauma. ISNA believes that this surgery is unnecessary, cosmetic and primarily "cultural" in its significance. It is of no benefit to the child, who suffers even more from the stigma and shame of having been surgically altered than she would have had her non-standard genitals been allowed to remain intact. Occasionally a female neonate will be so genitally virilized that she is given a male identity at birth and raised as a boy. It is important not to hide the circumstances of her biology from such a child, in order to avoid shame, stigma and confusion which results from secrecy. After the onset of puberty the child may want to explore the option, hopefully with the aid of loving parents and peer counseling, of having surgery to allow expression of either female or male sexuality. This is not a choice that should be forced prematurely, it is a personal choice to be made by a teenager about his/her body and about her/his choice of sexual identity and sexuality.

## **Swyer Syndrome**

\*Swyer Syndrome\* is also known as XY gonadal dysgenesis. In Swyer Syndrome, a person is born without functional gonads (sex glands). The gonads present in Swyer Syndrome are known as gonadal streaks. These are minimally developed gonad tissue present in place of testes or in place of ovaries. A child born with Swyer looks like a typical female. She will not develop most secondary sex characteristics without hormone replacement (medicines given by a doctor) because streak gonads are incapable of producing the sex hormones (both oestrogen and androgens) that bring about puberty.

## **Turner Syndrome**

The typical female karyotype ("sex" chromosome make-up) for females is 46,XX. This means that the typical female has 46 chromosomes including two that look like X's. People with Turner syndrome have only one X chromosome present and fully functional. This is sometimes referred to as 45,XO or 45,X karyotype. In a person with Turner Syndrome, female sex characteristics are usually present but underdeveloped compared to the typical female. The following signs are more common in women with Turner Syndrome than in the general population: short stature, lymphodema (swelling of hands and feet), broad chest and widely spaced nipples, low hairline, low-set ears, and infertility. However, Turner Syndrome shows up differently in different people—some signs associated with TS may be

more obvious in one woman than in the next. Mosaic Turner Syndrome can also occur. This is when some cells have two “sex” chromosomes (XX) but others only have one (X). A person can also have a mosaic in the form 46,XY/46X. Other mosaic types are also possible. When mosaic Turner Syndrome occurs, the person usually doesn’t have all the associated signs of TS, and may have other signs of intersex. For more information on Turner’s Syndrome visit: “Turner Syndrome at the National Institute of Child Health and Human Development”: <http://turners.nichd.nih.gov/> or “Turner Syndrome Society of the United States”: <http://www.turner-syndrome-us.org/>

### **5-alpha reductase deficiency**

Different from AIS, 5-ARDS occurs due to an autosomal defect (on a chromosome other than the X or Y chromosomes) and requires two altered genes, one from the father and one from the mother. 5-alpha-reductase is an enzyme that converts the weaker testosterone into the more potent *dihydrotestosterone (DHT)*. When this enzyme is deficient (*5-alpha-reductase deficiency*) the baby develops as a girl. However, at puberty testosterone production usually increases and is enough to cause virilization, so in such cases (where the child identifies strongly with a female role) it is advisable to perform a gonadectomy before puberty. On the other hand, there are cases where the child naturally migrates to a male role. Therefore, a conservative approach that incorporates listening to the child is imperative. 5-alpha reductase deficiency, unlike AIS, involves a defect on an autosome (not on a sex chromosome) and requires two mutated genes, one from the father and one from the mother.

### **What does ISNA recommend for children with intersex?**

After years of consultation with people with intersex conditions, their parents, their healthcare providers, and others, the following “Patient-Centered Model” is what ISNA recommends. \* Children with intersex, parents of those children, and adults with intersex should be treated in an open, shame-free, supportive, and honest way. They should consistently be told the truth (this includes providers being honest about uncertainty), and should be given copies of medical records as soon and as often as they ask for them. \* Children and adults with intersex, and their family members, should be provided with access to trained psychologists and social workers, especially when they are in distress (as some parents of newborns with intersex are). Parental distress should not be treated with “normalizing” surgery on children, nor should surgeons, endocrinologists, and other non-psycho-social specialists attempt to cover family’s counselling needs. \* Care providers should also attempt to connect children and adults with intersex and parents of children with intersex so that they can give each other peer support outside of the clinical setting. This helps validate their feelings and experiences. Peer support saves families and lives. \* Following diagnostic work-up, newborns with intersex should be given a “gender assignment” as boy or girl, depending on which of those genders the child is more likely to feel as she or he grows up. Note that gender assignment does not involve surgery; it involves assigning a label as boy or girl to a child. (Genital “normalizing” surgery does not create or cement a gender identity; it just takes tissue away that they patient may want later.) \* Medical procedures necessary to sustain the physical health of a child should be performed. Examples of these would be endocrinological treatment of a child with salt-wasting congenital hyperplasia, or surgery to provide a urinary drainage opening when a child is born without one. \* Surgeries done to make the genitals look “more normal” should not be performed until a child is mature enough to make an informed decision for herself or himself. Before the patient makes a decision, she or he

should be introduced to patients who have and have not had the surgery. Once she or he is fully informed, she or he should be provided access to a patient-centered surgeon. Does this mean ISNA recommends "doing nothing"? Not at all. Please re-read the above, and if you'd like more information about how our recommendations differ from the traditional "concealment-centered" model, click here to check out our chart called "*Shifting the Paradigm of Intersex Treatment*".

### **Does ISNA think children with intersex should be raised without a gender, or in a third gender?**

No, and for the record, we've never advocated this. We certainly would like to see people become less freaked-out by people who don't fit sex and gender cultural norms. But there are at least two problems with trying to raise kids in a "third gender." First, how would we decide who would count in the "third gender"? How would we decide where to cut off the category of male and begin the category of intersex, or, on the other side of the spectrum, where to cut off the category of intersex to begin the category of female? Second, and much more importantly, we are trying to make the world a safe place for intersex kids, and we don't think labelling them with a gender category that in essence doesn't exist would help them. ISNA recognizes that it can be damned hard to be intersex, or to have an intersex child. That's why we exist. That's why we don't advocate doing nothing. What we do advocate is providing parents of intersex newborns--and within a couple of years, intersex children themselves--honest and accurate information about intersex, psychological counselling by professionals who are not intersex-phobic, medical help for any real medical problems, and especially referrals to other people dealing with the same issues. Time and again researchers have found that, no matter what the condition--being gay, dealing with a serious disease--peer support, even if informal, saves families and lives.

### **What's wrong with the way intersex has traditionally been treated?**

In the 1950s, a team of medical specialists at Johns Hopkins University developed what has come to be called the "optimum gender of rearing" system for treating children with intersex. The notion was that the main thing you had to do in cases of intersex was to get the gender assignment settled early, so kids would grow up to be good (believable and straight) girls and boys. Under the theoretic leadership of psychologist John Money, the Hopkins team believed that gender was all about nurture--that you could make any child into a "real" girl or boy if you made their bodies look right early (before about 18 months of age), and made them and their parents believe the gender assignment. Though the Hopkins team wrote early on that children should be told the truth about their intersex histories in age-appropriate ways, in practice many medical care providers lied to patients or actively withheld medical history information from them. Medical textbooks frequently gave doctors advice about how to lie to patients with intersex. For examples of people who were lied to and deceived, check out "Hermaphrodites Speak":/videos/hermaphrodites\_speak and "Chrysalis":/books/chrysalis at [www.isna.org](http://www.isna.org). As the "Hopkins model" spread throughout the developed world, surgeons performed cosmetic genital surgeries on intersex children without their consent, believing this was necessary and efficacious. Endocrinologists, meanwhile, manipulated patients' hormones to try to get the bodies of patients to do what they thought was necessary not just for physical health, but for psycho-social health (i.e., getting the body to look sexually "normal"). So what was wrong with this model? To start with, lying to patients is not only unethical, it is bad medicine. Patients who were lied to figured that much out, and often stopped getting medical care they needed to stay healthy. (For example, some stopped taking hormone replacement therapy--critical after gonadectomy--and wound up with

life-threatening osteoporosis at an early age.) They also suffered psychological harm from these practices, because they got the message that they were so freakish even their doctors could not speak the truth of their bodies to them. (A lot of doctors still have not told their present and former patients the name of their conditions. Some still withhold medical records from patients and from parents/guardians of minor children.)

Second, the system was and is literally sexist: that is, it treats children thought to be girls differently than children thought to be boys. In this approach (still going on at Hopkins so far as we can tell), doctors' primary concern for children thought to be girls is preservation of fertility (not sexual sensation), and for children thought to be boys, size and function of the phallus. Third, the "standards" used for genital anatomy have been arbitrary and illogical. For example, under the "optimum gender of rearing" model, boys born with penises doctors considered small were made into girls--even though other doctors believed, and showed they could be raised as boys without castration, genital surgery, and hormone replacement, (for evidence boys with small penises do well without sex re-assignment, see Justine M. Reilly and C. R. J. Woodhouse, "Small Penis and the Male Sexual Role," *Journal of Urology*, 142 (1989): 569-571). Girls with clitorises their doctors think are "too big" still find themselves in operating theatres with surgeons cutting away at their healthy genital tissue. Paradoxically, though all medical experts agree the identification of intersex anatomy at birth is primarily a psycho-social (not medical) concern, it is still treated almost exclusively with surgery. Parental distress is treated with the child being sent off to surgery. This is not an appropriate form of care for parents or children. (See Edmund G. Howe, "Intersexuality: What Should Care Providers Do Now," *Journal of Clinical Ethics*, 9, 4 (Winter 1998): 337-344). There is no evidence that children who grow up with intersex genitals are worse off psychologically than those who are altered. In fact, there is evidence that children who grow up with intersex genitals do well psychologically. In other words, these surgeries happen before the age of assent or consent without real cause. "Ambiguous" genitalia are not diseased, nor do they cause disease; they just look funny to some people. There is substantial evidence that people who have been treated under the "optimum gender of rearing" model have suffered harm, psychological and physical. This does not mean doctors intended to harm their patients; far from it. But good intentions are inadequate reasons to maintain a practice that has shown to be unethical and unscientific. Finally, parents consenting to intersex surgeries do not appear to be fully informed about the available evidence, about alternatives available to them, about the risks associated with surgeries, or about the theoretical problems underlying the "optimum gender of rearing" approach. For example, they are typically not told the evidence that gender identity may emerge to an important degree from prenatal hormonal actions on the brain--and thus, that you can't "make" a child maintain a particular gender identity in the long term by doing surgery on him or her in infancy. On this point, see William G. Reiner and John P. Gearhart, "Discordant sexual identity in some genetic males with cloacal exstrophy assigned to female sex at birth," *New England Journal of Medicine*, 350, 4 (Jan. 22, 2004): 333-341.

### **What do doctors do now when they encounter a patient with intersex?**

So far as we can tell, most medical centres still practice the "concealment-centred model of care" that grew out of Hopkins' "optimum gender of rearing". We still hear many reports of "normalizing" (medically unnecessary) genital surgeries and hormone treatments that were not consented to by the patient, and of adult patients and parents of minors being denied medical records. What we heard at the "American Academy of Paediatrics meeting":/articles/aap\_urology\_2004 leads

us to think doctors are fairly aware of the controversy surrounding intersex treatment, but are still taking the basic approach of "cut now, maybe ask about quality of life later." We are hoping to obtain funding soon for an Audit of Care project which would allow us to visit several medical centres claiming to use a more "patient-centered" model of care. We hope to find out that they are, in fact, doing progressive work, because if they are, we can hold them up as models of leadership.

### **How come many people have never heard of intersex?**

For decades, doctors have thought it necessary to treat intersex with a "concealment-centered approach", one that features downplaying intersex as much as possible, even to the point of "lying to patients. A lot of people in our culture also had no interest in hearing that sex doesn't come in two simple flavours. But that has been changing since "Cheryl Chase" founded ISNA in 1993. For over a decade we at ISNA have been successfully working to de-stigmatize intersex and to let people know that folks with intersex are all around. ISNA representatives have appeared in dozens of local, national, and international television and radio programs, and in virtually all major newspapers and magazines. We estimate conservatively that we've reached 30 million people since 1993. People who are intersex will tell you that the primary thing they've been harmed by is induced shame about their intersex. The best way to reduce shame (and thereby reduce harm to individuals and families coping with intersex) is to talk openly and honestly about intersex. If you've learned something new at our site today, tell someone else about it!

### **Is a person who is intersex a hermaphrodite?**

No. The mythological term "hermaphrodite" implies that a person is both fully male and fully female. This is a physiologic impossibility. The words "hermaphrodite" and "pseudo-hermaphrodite" are stigmatising and misleading words. Unfortunately, some medical personnel still use them to refer to people with certain intersex conditions, because they still subscribe to an outdated nomenclature that uses gonadal anatomy as the basis of sex classification. In a paper titled "Changing the Nomenclature/Taxonomy for Intersex: A Scientific and Clinical Rationale":/node/979, five ISNA-associated experts recommend that all terms based on the root "hermaphrodite" be abandoned because they are scientifically specious and clinically problematic. The terms fail to reflect modern scientific understandings of intersex conditions, confuse clinicians, harm patients, and panic parents. We think it is much better for everyone involved when specific condition names are used in medical research and practice.

While some intersex people seek to reclaim the word "hermaphrodite" with pride to "reference themselves" (much like the words "dyke" and "queer" have been reclaimed by LBGT people), we've learned over the years it is best generally avoided, since the political subtlety is lost on a lot of people.

### **Does having a Y chromosome make someone a man?**

A lot of unintended harm happens when people assume a Y chromosome makes a person a boy or a man and the lack of a Y chromosome makes a person a girl or a woman. For example, one physician educator on our "Medical Advisory Board" had the challenging experience of trying to calm a 23-year-old patient who had just been told by a resident that she was "really a man" because the resident had diagnosed the patient as having a Y chromosome and complete "androgen insensitivity syndrome" (CAIS). It is true that in typical male development, the SRY gene on the tip of the Y chromosome helps to send the embryo down the masculine

pathway. But more than the SRY is needed for sex determination and differentiation; for example, women with CAIS have the SRY gene but lack androgen receptors. In terms of hormone effects on their bodies (including their brains), women with CAIS have had much less "masculinization" than the average 46,XX woman because their cells do not respond to androgens. Moreover, the SRY gene can be translocated onto an X chromosome (so that a 46,XX person may develop along a typical masculine pathway), and there are dozens of genes on chromosomes other than the X and the Y that contribute to sexual differentiation. And beyond the genes, a person's sex development can be significantly influenced by environmental factors (including the maternal uterine environment in which the foetus developed). So it is simply incorrect to think that you can tell a person's sex just looking at whether he or she has a Y chromosome. Want to know more? The following comes from ISNA's Medical Advisory Board member Dr. Charmian Quigley: SRY, discovered in 1989, is a small gene located at the tip of the short arm of the Y chromosome. So what does it do? Actually, like all genes, it does nothing except to act as a blueprint for a protein. In this case, the protein of the same name does funky things to DNA, like bending it and unwinding the 2 strands, so that other proteins can get in and attach themselves to other genes that are then turned on. So how did this gene get its reputation (and its name) as the "sex determining" gene? As is pretty common in the world of genetics, this was because of some errant mice. Researchers in England took a laboratory-made copy of this gene and inserted it artificially into a female (XX) mouse embryo at a very early stage of development. The mouse was "converted" from female to male, so the gene must have been responsible - right? Well, maybe not. A few years later, a similar gene was found on human chromosome 17. When the important part of this gene was inserted into a female mouse embryo, the same thing happened. Voila! A male. So now we have 2 genes that can turn a female into a male, and one of them is not located on the Y chromosome! How can that be? It turns out that SRY is probably just a facilitator that allows a more critical gene (or genes) to function, by blocking the action of another opposing factor. Can the magic of genetics do the opposite - turn a male into a female? Indeed it can. A gene on the X chromosome (the chromosome one typically associates with "femaleness") called DAX1 when present in double copy in a male (XY) mouse, turns it into a female. So now we have genes on the Y that can turn females with XX chromosomes into males and genes on the X that can turn males with XY chromosomes into females. . . wow! Maleness and femaleness are NOT determined by having an X or a Y, since switching a couple of genes around can turn things upside down. In fact, there's a whole lot more to maleness and femaleness than X or Y chromosomes. About 1 in 20,000 men has no Y chromosome, instead having 2 Xs. This means that in the United States there are about 7,500 men without a Y chromosome. The equivalent situation - females who have XY instead of XX chromosomes - can occur for a variety of reasons and overall is similar in frequency. For these 15,000 or more individuals in the US (and who knows how many worldwide), their chromosomes are irrelevant. It is the total complement of their genes along with their life experiences (physical, mental, social) that makes them who they are (or any of us, for that matter). The last time I counted, there were at least 30 genes that have been found to have important roles in the development of sex in either humans or mice. Of these 30 or so genes 3 are located on the X chromosome, 1 on the Y chromosome and the rest are on other chromosomes, called autosomes (on chromosomes 1, 2, 3, 4, 7, 8, 9, 10, 11, 12, 17, 19). In light of this, sex should be considered not a product of our chromosomes, but rather, a product of our total genetic makeup, and of the functions of these genes during development.

### **Is intersex the same as "ambiguous genitalia"?**

No, saying someone has an intersex condition isn't the same as saying she or he was born with "ambiguous genitalia," because some people with intersex conditions have genitalia that look pretty typically masculine or feminine. So, for example, girls born with XY chromosomes and complete androgen insensitivity syndrome have genitals that look pretty typically female. And some children born with XX chromosomes and congenital adrenal hyperplasia are born with genitals that look thoroughly male. Yet nearly all medical professionals agree that these kinds of conditions are intersex. Why do we put the term "ambiguous genitalia" in quotation marks? We don't particularly like the term since, as our "Medical Advisory Board" member Dr. William Reiner likes to point out, no child thinks his or her own genitals are "ambiguous." They're just their genitals. It's the grown-ups who are feeling ambiguous.

### **What's the difference between being transgender or transsexual and having an intersex condition?**

People who identify as transgender or transsexual are usually people who are born with typical male or female anatomies but feel as though they've been born into the "wrong body." For example, a person who identifies as transgender or transsexual may have typical female anatomy but feel like a male and seek to become male by taking hormones or electing to have sex reassignment surgeries. People who have intersex conditions have anatomy that is not considered typically male or female. Most people with intersex conditions come to medical attention because doctors or parents notice something unusual about their bodies. In contrast, people who are transgendered have an internal experience of gender identity that is different from most people. Many people confuse transgender and transsexual people with people with intersex conditions because they see two groups of people who would like to choose their own gender identity and sometimes those choices require hormonal treatments and/or surgery. These are similarities. It's also true, albeit rare, that some people who have intersex conditions also decide to change genders at some point in their life, so some people with intersex conditions might also identify themselves as transgender or transsexual. In spite of these similarities, these two groups should not be and cannot be thought of as one. The truth is that the vast majority of people with intersex conditions identify as male or female rather than transgender or transsexual. Thus, where all people who identify as transgender or transsexual experience problems with their gender identity, only a small portion of intersex people experience these problems. It's also important to understand the differences between these two groups because in spite of some similarities they face many different struggles, including different forms of discrimination. The differences between transgender and transsexual and intersex have been understood by lawmakers in countries such as Australia where "lawmakers have publicly acknowledged that people with intersex conditions have distinct needs from people who identify as transgender or transsexual" [http://home.vicnet.net.au/~aissg/transgender\\_and\\_intersex.htm](http://home.vicnet.net.au/~aissg/transgender_and_intersex.htm). People who identify as transgender or transsexual also face discrimination and deserve equality. We also believe that people with intersex conditions and folks who identify as transgender or transsexual can and should continue to work together on human rights issues; however, there are important differences to keep in mind so that both groups can work toward a better future.

## Why Doesn't ISNA Want to Eradicate Gender?

We're often asked why ISNA doesn't forcefully advocate for a genderless society. Many times, these questions come from people with a genuine interest in gender studies and educating people about intersex. The truth is that we share lots of common ground with people in the humanities and/or activist communities who have fought long and hard to insure that the voices of marginalised people are heard. When women of colour told feminists that their lives weren't reflected in theories that assumed white experience to be universal, scholars listened. When queer people came forward to say that theories of gender that neglected sexuality often fell short of capturing the realities of their lives, scholars listened. Without a doubt, scholars have a rich history of taking the voices of marginalized people seriously and changing their theories and practices accordingly, and now ISNA asks that scholars listen to what people with intersex conditions have to say--even if it might not be what they'd like to hear. At ISNA, we've learned that many intersex people are perfectly comfortable adopting either a male or female gender identity and are not seeking a genderless society or to label themselves as a member of a third gender class. Although it's true that the urge to perform surgeries on intersex children's sex anatomies is sometimes born out of the belief that children must have sex anatomies that are clearly male or female in order to be comfortable in either a male or female gender (and this is clearly a harmful belief born out of antiquated notions about gender identity corresponding directly to genital anatomy), the idea of raising a child as a boy or girl isn't what most adults with intersex conditions point to as their main problem. In fact, many of the people with intersex we know--both those subjected to early surgeries and those who escaped surgery--very happily accepted a gender assignment of male or female (either the one given them at birth or one they chose later for themselves later in life). Instead, adults with intersex conditions who underwent genital surgeries at early ages most often cite those early genital surgeries and the lies and shame surrounding those procedures as their source of pain. Later in life, like many people with typical anatomies, intersex people take pleasure in what some gender scholars (like Judith Butler) might call doing their gender. Thus, intersex people don't tell us that the very concept of gender is oppressive to them. Instead, it's the childhood surgeries performed on them and the accompanying lies and shame that are problematic. Again, many of these surgeries are performed with the belief that these procedures will help a child settle into a gendered world, but that doesn't mean the whole system of gender must fall in order for people with intersex conditions to live happy, fulfilling lives. It simply means that these surgeries and the shame that surrounds them are an unfortunate instantiation of problematic gender norms and we should work on ending unwanted surgeries and stigma. There are, of course, some people with intersex conditions who identify as a third gender or gender queer--just as there are some people with completely typical sex anatomies who don't identify as strictly male or female. Our aim at ISNA isn't to undermine these people's goals, or to suggest that people who identify as a third gender don't exist or don't matter, or to suggest that everyone must adopt a gender. Rather, we hope to end painful and unnecessary childhood surgeries that rob people of corporeal autonomy and sexual function because everyone--regardless of gender identity--deserves that. And we hope to end the shame and secrecy that cause so much pain for so many people with intersex conditions. We hope that scholars, particularly those invested in helping members of marginalised groups gain a voice in conversations about themselves, will take seriously the concerns about surgery, secrecy, and shame raised by intersex people and understand that ISNA and the majority of its constituency don't necessarily share the goal of eradicating the very notion of gender.

## **How can you assign a gender (boy or girl) without surgery?**

When you assign a child a gender as boy or girl, what you're doing is labelling them a boy or girl. That's it. You don't need a surgeon for that. But how do you pick a child's gender if she or he is intersex? The child is assigned a gender as boy or girl after tests (hormonal, genetic, radiological) have been done and the parents have consulted with the doctors on which gender the child is more likely to feel as she or he grows up. We know, for example, that the vast majority of children with complete androgen insensitivity syndrome grow up to feel female, and that many children with cloacal exstrophy and XY chromosomes will grow up to feel male. Why shouldn't children with intersex be raised in a "third gender"? We advocate assigning a boy or girl gender because intersex is not, and will never be, a "discrete biological category any more than male or female is, and because assigning an "intersex" gender would unnecessarily traumatize the child. In cases of intersex, doctors and parents need to recognise, however, that gender assignment of infants with intersex conditions as boy or girl, as with assignment of any infant, is preliminary. Any child--intersex or not--may decide later in life that she or he was given the wrong gender assignment; but children with certain intersex conditions have significantly higher rates of gender transition than the general population, with or without treatment. That is a crucial reason why medically unnecessary surgeries should not be done without the patient's consent; the child with an intersex condition may later want genitals (either the ones they were born with or surgically constructed anatomy) different than what the doctors would have chosen. Surgically constructed genitals are extremely difficult if not impossible to "undo," and children altered at birth or in infancy are largely stuck with what doctors give them.

## **What evidence is there that you can grow up psychologically healthy with intersex genitals (without "normalising" surgeries)?**

A lot of people think that doctors do "normalising" surgeries on infants and children with intersex because, if they didn't, those children would grow up to be very damaged psychologically. In fact, there's virtually no documented evidence for that. Yup, you heard us right: there's virtually no evidence of people with "uncorrected" intersex genitals suffering increased rates of psychological illness or social ostracisation. In fact, we do have lots of evidence that people who grow up with "uncorrected" intersex genitals do OK. Here's some of it: A 1937 book, *Genital Abnormalities, Hermaphroditism, and Related Adrenal Diseases* (Baltimore: Williams and Wilkins) recorded the lives of dozens of adults with "sexual ambiguity," very few of whom were seeking "corrective" surgery. Most were fine with their physical differences. Ironically, the author of that book, Hugh Hampton Young, was a famous surgeon at Johns Hopkins University, the very place that ultimately launched the "optimum gender of rearing" that insisted you had to do "normalising" surgery on very young children. If the Hopkins team had read Young's work, maybe they would have thought twice about how necessary infant genital surgeries really are. But probably not. Because the very man at Hopkins leading the charge for infant surgeries had himself shown, through extensive research, that people with "uncorrected" intersex have lower rates of psychopathology than the general population. That man was John Money. For his Ph.D. dissertation in psychology at Harvard University in 1952, Money researched hundreds of people with intersex. And yes, he found that they had lower rates of psychopathology than the general population. His theory was that they learned good coping skills. You can read about this in Money's dissertation (never published, but you can order it from Harvard University's Widener Library for about \$100) or read the summary of it in John Colapinto's book, "As Nature Made Him". In spite of the fact that the

Hopkins surgery-based system for treating intersex "spread all over the world", some people slipped through, growing up with intersex genitals intact. All the ones we know about did OK. In our video, "Hermaphrodites Speak!":/videos/hermaphrodites\_speak, Hida Vioria tells about how she is glad she never had "corrective" surgery. Hida also testified at the "San Francisco Human Rights Commission's open hearing on intersex":/videos/sf\_hrc\_hearing. Hale Hawbecker--whose parents went against medical advice to sex re-assign him at birth--reports the same in chapter 11 of "Intersex in the Age of Ethics":/books/ageofethics. Also take a look at chapters 7 and 9 in that same book for autobiographies of two women who grew up with large clitorises. As early as 1989, a peer-reviewed study published in the *Journal of Urology* showed that boys with "micropenis" could do well if raised without surgery and with honesty. Reilly (whose married name is Justine Schober and who is now member of our "Medical Advisory Board) and Woodhouse "interviewed and examined 20 patients with the primary diagnosis of micropenis in infancy" and concluded that "[A] small penis does not preclude normal male role and a micropenis or microphallus alone should not dictate a female gender reassignment in infancy." More particularly, these doctors found that when parents "were well counselled about diagnosis they reflected an attitude of concern but not anxiety about the problem, and they did not convey anxiety to their children. They were honest and explained problems to the child and encouraged normality in behaviour. We believe that this is the attitude that allows these children to approach their peers with confidence." (See Justine M. Reilly and C. R. J. Woodhouse, "Small Penis and the Male Sexual Role," *Journal of Urology*, 142 (1989): 569-571.)

This, again, accords with what we know happened to people before the era of enforced infant "normalising" surgeries. In "Hermaphrodites and the Medical Invention of Sex":/books/medicalinvention, historian Alice Dreger documents many cases of French and British people who, before the age of surgical "correction", fared well socially. Some of them sought advice and information from medical doctors, but very few sought "corrective" surgeries. There has always been sex variation, and there have not been widespread suicides of or assaults on people with variations that could easily be labelled intersex. In his 1949 book, *Human Sex Anatomy* (Baltimore: The Williams and Wilkins Company), Robert Latou Dickenson demonstrated how adult women and men have widely varied forms of genital anatomy. So, what's the evidence that children with intersex actually need "normalising" genital surgeries--surgeries that risk their health, continence, fertility, sensation, and life? We're waiting to see it.

### **What's ISNA's position on surgery?**

Many people who know of ISNA's work mistakenly think we are "anti-surgery." Not at all! Like all sane people, we believe it is appropriate to have competent surgeons perform operations necessary to resolve a life-threatening metabolic crisis. For example, if a child is born without a urinary opening, the child needs surgery to create a urinary opening. If a child has active gonadal cancer, the cancer should be treated immediately. What about other kinds of surgeries in cases of intersex? We believe that competent patients should be allowed to get the surgeries they want after they have been fully informed of the risks and benefits (and the evidence, or lack thereof, for both). They should be given access to expert, humane surgeons, as well as peer support before and after their procedures. What we object to are elective surgeries done on people (usually children) without their informed consent. Such surgeries subject patients to unnecessary harm and risk.

## **Are there medical risks associated with intersex conditions?**

### **Congenital Adrenal Hyperplasia (CAH) medical risks**

"Congenital Adrenal Hyperplasia" (CAH) is the only one of all the various causes of intersex that can actually cause a medical emergency. In fact, before CAH was well understood, it was thought to occur much more frequently in girls. Now we know that this really reflects the fact that newborn girls with genital ambiguity often came quickly to medical attention, whereas CAH boys (who don't have an intersex appearance) often died without being diagnosed. CAH occurs equally often in XX and XY individuals. People with CAH (both XY individuals, who are not intersexed, and XX individuals, who may be intersexed) have problems making their own cortisone, a hormone which helps the body respond to stress (like a cold, or a broken bone, or severe trauma in a bad automobile accident, or undergoing surgery). This problem can be addressed by administering cortisone, but only if medical professionals know that the person has CAH. For this reason, some CAH people wear Medic Alert bracelets. Another problem that can happen is called "Salt wasting." Some people with CAH (whether intersexed or not) don't produce the right levels of hormones that control salt in the body. These people can get very sick, and even die, without medication to correct their body's salt level. Many people with "salt wasting" CAH also say that they crave salt, and feel better when they eat very salty food. Any child who has XX chromosomes and CAH, and is raised as a boy may experience feminizing puberty and menstruation. We believe that in such a case, the child deserves a clear explanation of his own medical condition, and a choice about medical interventions. A mental health specialist, working with the child and the parents, should determine the child's gender identity. If the child's identity is female, she may wish to have psychosocial support to change her social sex to female. She may also wish cosmetic genital surgery to make her genitals look more female. If the child's gender identity is male, he may wish to prevent menstruation from occurring. If it is difficult to determine the child's gender identity or wishes, puberty can be temporarily delayed with the drug "Lupron" <http://www.lupron.com/>. This is not a permanent solution, but a delaying tactic. If the child's gender identity is male, he may choose to have his ovaries removed to prevent further feminisation. As reproductive technology advances, consideration should be given to preserving gonadal tissue for future assisted fertility.

### **Gonadal tumours**

In general, gonadal tumours are unlikely in the absence of a Y chromosome or Y genes which may be present on the X chromosome. When there is a Y chromosome or Y genes are surmised to be present, the gonads are at elevated risk, and should be carefully monitored. Because the risk is slight before early adulthood, gonadectomy should not be imposed on infants. It should be delayed until the patient can weigh the options and choose for her/himself. Functioning gonads, even partially functioning gonads, are a big advantage over hormone replacement therapy. The patient must be allowed to weigh the risks, talk with other patients about their experiences, and choose what is best for her/himself. Note, though, that it is critical to remove partially functioning testes before puberty from an intersexual who identifies as female and wishes her body not to virilize.

### **Osteoporosis**

Testosterone or oestrogen are necessary to maintain healthy adult bones. If you were born without functioning gonads (ovaries or testes), or if your gonads have been removed, you should be under an endocrinologist's care and maintain hormone replacement therapy for life. Many people with intersex conditions,

having developed a distrust or aversion for medical people, avoid medical care and drop hormone replacement therapy which was prescribed during puberty. This can result in extreme osteoporosis (brittle bones). Osteoporosis worsens silently, but at advanced stages it can destroy your quality of life. Persons with advanced osteoporosis are vulnerable to frequent bone fractures, especially of the spine, hip, and wrist. These fractures can be caused by a small amount of force, and are extremely painful and debilitating. Each spine fracture may put you flat on your back for one to two months. If you have been without gonads or hormone replacement therapy for years, it is vital to get a bone density scan performed, to evaluate the condition of your bones (a simple, non-invasive procedure using a specialized x-ray machine), and to seek the advice of an endocrinologist in order to establish a regimen of hormone replacement therapy that works for you. If you have had bad experience in the past with hormones, we encourage you to find an endocrinologist who will work with you to adjust the mix and schedule of hormones until you find what works. If your bone density is low, your endocrinologist will probably recommend calcium supplements and weight-bearing exercise (not swimming!) to maintain density. If your bone density scan is performed on a DEXA machine, make certain to do any follow-up scans on the same machine, and with the same reader. A number of drugs currently in the biomedical news may prove useful for rebuilding lost bone density. If your bone density is low, check in with a qualified specialist regularly for the latest information. The danger of osteoporosis is considerably worse for people with intersex conditions than for post-menopausal women, because they will be without hormones for many decades.

### **How can I get my old medical records?**

If you know that you were "identified as having an intersex condition":/node/726 as an infant, you may want to try to get your old medical records. But if you do know that you have an intersex condition, we encourage you to try to get copies of your medical records. Many of us have found it invaluable to obtain copies of our medical records, especially those of our births or early genital surgeries. The medical information and the emotional confirmation of what was done to us as children and how we were evaluated by medical personnel helps us to heal. With this information we can better know ourselves and understand who we are. In Australia, if an institution or a physician has your records, you have a legal right to a copy of those records, no matter how old. Until recently, hospitals rarely discarded old records. Sometimes they are moved into warehouses or onto microfilm. Now there is a trend to throw out old records. You may be able to obtain your records by having a physician request them. If your records are not easily found and your first request is refused or ignored, you may be able to enlist the help of a records clerk. Try telling your story; if you can enlist his or her sympathy, the clerk may be willing to look harder for records misfiled or stored in a warehouse or on microfilm. Showing up in person at the hospital or office may help.

### **What do intersex and the same-sex marriage debate have to do with each other?**

A lot, as it turns out. People who are proponents of prohibitions against "same sex" marriage think it is easy to figure out who is "same sex" and who is "opposite sex." Not so. Check out the first section above called "What is intersex?" to learn more about how it isn't clear, in practice, where the category of "male" should end and "intersex" begin, or "intersex" end and "female" begin. Lots of people with intersex that we know are legally married. What will happen to them if we end up with simplistic notions of sex? And lots of people with intersex we know can't get legally married, because some doctor decided for them which sex they would count as

forever more. Why should a doctor get to decide who you can grow up to marry? To read more about this, go to: \* Alice Dreger's blog, "What Intersex Does to the Gay Marriage Debate":/node/670 \*, A "report on a clash":

[http://www.gaycitynews.com/gcn\\_421/panelokstrangendered.html](http://www.gaycitynews.com/gcn_421/panelokstrangendered.html) between the Department of Homeland Security and the Board of Immigration Appeals \*, Stephen Jacquier's UPI commentary, "Gender & the Marriage Debate":  
<http://www.washingtontimes.com/upi-breaking/20041117-092204-4819r.htm>

### **Who was David Reimer (also, sadly, known as "John/Joan")?**

David Reimer was born an identical (non-intersex) twin boy in 1965. At the age of 8 months, David and his brother each had a minor medical problem involving his penis, and a doctor decided to treat the problem with circumcision. The doctor botched the circumcision on David, using an inappropriate method and accidentally burning off virtually all of David's penis. At the advice of psychologist John Money at Johns Hopkins University, David's parents agreed to have him "sex reassigned" and made into a girl via surgical, hormonal, and psychological treatments--i.e., via "the system Money advocated for intersex children". For many years, John Money claimed that David (known in the interim as "Brenda") turned out to be a "real" girl with a female gender identity. Money used this case to bolster "his approach to intersex" --the approach that is still used throughout much of the U.S. and developed world--one that relies on the assumption that gender identity is all about nurture (upbringing), not nature (inborn traits), and that gender assignment is the key to treating all children with atypical sex anatomies. As it turns out, Money was lying. He knew Brenda was never happy as a girl, and he knew that as soon as David found out what happened to him, David reassumed the social identity of a boy. The case of David Reimer has been used by the proponents of the "gender is inborn" (nature) theory as proof that they are right. We like to point out that what the story of David Reimer "teaches us most clearly" is how much people are harmed by being lied to and treated in inhumane ways. We don't think we can ever predict, with absolute certainty, what gender identity a person will grow up to have. What we can predict with a good degree of certainty is that children who are treated with shame, secrecy, and lies will suffer at the hands of medical providers who may think they have the best of intentions and the best of theories. To read more about David Reimer, see: \* "David Reimer: The Boy who Lived as a Girl":  
<http://www.cbc.ca/news/background/reimer/> at Canada's CBC News.  
John Colapinto's original article about David Reimer in "Rolling Stone":  
<http://www.pfc.org.uk/news/1998/johnjoan.htm>  
"The Death of David Reimer: A tale of sex, science, and abuse" :  
<http://www.reason.com/links/links052404.shtml>  
by Jesse Walker, at Reason.

### **What's the history behind the intersex rights movement?**

Beginning in the late nineteenth century, medicine became the primary means for dealing with intersex. Before then, the vast majority of people with intersex conditions went unnoticed by legal, religious, or medical establishments and only a few cases per year came to the attention of authorities. Presumably other people with so-called "abnormal" sex anatomies lived average lives, either because their anatomical variance was undetectable or was not considered especially important. When a newborn had a high degree of genital ambiguity, midwives, grandmothers, and other local elders appear to have assigned the sex. (In terms of sexual orientation, all people were expected to then have sexual relations solely with those who had been identified as the "opposite" sex; in many places, violation of this rule was punishable by violent, sometimes fatal means.) However, by the late 1800s, through gynaecological sciences and numerous wartime military medical

examinations, doctors gained a much better sense that "abnormal" sex anatomies were actually quite common. Indeed, late-nineteenth century medical men began reporting dozens of cases a year of "hermaphroditism" and "pseudo-hermaphroditism." Because most medical experts were politically conservative and wanted to keep sex borders clearly defined to combat open homosexuality and the rise of feminism, intersex caused them notable stress. (The conflation of sex, sexual orientation, and gender expression becomes clear in the 1890s use of the term "psychic hermaphroditism" to refer to gay men, and in the common "scientific" claim that university education physically "masculinised" women.) Therefore, biomedical specialists devised a system that would label everyone "truly male" or "truly female," regardless of the extent and natural reality of sexual blending. Medical doctors created an "arbitrary standard based on gonadal tissue", which persists in most medical texts today. A person with non-standard sex anatomy and ovaries is seen as a "female pseudo-hermaphrodite"; a person with non-standard sex anatomy and testes, is seen as a "male pseudo-hermaphrodite"; and if a person has ovarian and testicular tissue, they are seen as a "true hermaphrodite." Given the technological limitations of the time, Victorian doctors liked this system because they couldn't easily diagnose "true hermaphroditism" in living people; as a consequence, for the most part the only "true hermaphrodites" were dead and dissected people and the only medical information about intersex came from posthumous examinations. All other people thought to be intersex, including pseudo-hermaphroditic, were labelled "truly male" or "truly female" and told to act socially and sexually normative in their assigned gender. However, with improved medical techniques and increased access to healthcare, many more people were being diagnosed with a biological "true sex" that made no sense socially. (In the 1910s as today, women with androgen insensitivity couldn't practically be labelled men just because they had testes.) And, in a bi-polar gender paradigm, there was no simple social category for those diagnosed with "true hermaphroditism." So, by the 1920s, experts treating intersex developed a notion of gender (social role) separated from biological sex. And they began to more actively offer surgical "corrections" to bring the biological sex into line with the assigned gender. So the theoretical approaches and surgical techniques evolved bit by bit, though motivation remained the same: "keeping sex categories distinct and numbering only two". It should be noted that, in the last half of the nineteenth century, a small percentage of patients with intersex had started to ask for, and some surgeons had started to offer surgical reconstruction of large clitorises, small vaginas, and hypospadiac penises. With the exception of the rare clitorectomy performed on a child because she had a large clitoris, most of the genital surgeries performed for cosmetic reasons in the nineteenth century were performed on adults at their request. Both patients and surgeons generally avoided elective surgery for reasons of safety. There is also reason to believe that most people with intersex were socially healthy without surgery; they did not disproportionately live as hermits or attempt suicide. Psychologist John Money studied adults with intersex and found--before the era of standard cosmetic surgical intervention for intersex--that they enjoyed a lower rate of psychopathology than the general population. Nevertheless, like many other realms of biology, sexuality, and psychology, intersex increasingly became the purview of medicine. For a small number of people with intersex--namely those at risk for gonadal cancers and salt-wasting--the medicalisation of intersex probably improved their health, sometimes even saving their lives. Nevertheless, most of the treatment of intersex was motivated not by metabolic health concerns, but by psychosocial concerns; as in the 1890s, by the 1950s, intersex was viewed primarily as a psychosocial problem that mixed sex categories in socially uncomfortable ways. In the 1950s, Johns Hopkins University created a team and became the first medical centre to offer an organised multi-

disciplinary approach to intersex, one that sought to essentially eliminate intersex in early childhood. The approach developed there came to be known as the "optimum gender of rearing" model. The basic idea was that each child's potential for a "normal" gender identity should be maximised by making each child's body, upbringing, and mind align as much as possible. Because of the belief that it was harder to surgically engineer a boy than a girl, most children with intersex were made as feminine as possible, utilizing surgery, endocrinology, and psychology. A "successful" patient was one judged to be stable and "normal" (i.e., heterosexual) in the assigned gender. (In an era of vice squads raiding gay bars, it is not surprising that homosexuality appeared to most of these professionals an untenable identity.) Though the early texts from this team advocated truth-telling and consistent psychological support, in practice many patients were deceived and given minimal psychological support. As in most of medicine, doctors made the decisions for patients. There was little investment in the ideas of informed consent or of studying outcomes in a systematic way. (It wasn't until the 1960s that medical professionals began aggressively looking for cases of intersex.) Psychologist John Money provided theoretical support for the Hopkins model, arguing that gender identity was largely mutable early in life, that nurture was more important than nature. His chief support for this claim came from a famous case known as "John/Joan." The person at the center, "David Reimer", was born an identical twin (not intersex) boy in 1965. While performing a circumcision, a doctor accidentally burned off eight-month-old David's penis. David's parents consulted with the team at Hopkins, and Money recommended they change the sex and gender of their child and raise David as a girl. For decades Money erroneously touted a successful outcome because the child reportedly had become a normal, female-identified, heterosexual girl. "The truth" was that David had never felt fully female. Indeed, when his parents told him the truth about what happened to him, he immediately re-assumed the gender role of a man. It is unknown why Money--who in 1953 had found a relatively low rate of psychopathology among adults with intersex--thought people with intersex needed to have their sexes and genders surgically and socially engineered to be psychologically healthy. It is better understood why people did not question Money's controversial theory of nurture-over-nature approach. Surgeons and psychologists liked the theory because it signified that they were providing necessary, good care to "abnormal" children. Feminists liked the theory because they preferred the idea that gender - and therefore gender norms - were socially constructed and malleable. Parents probably liked it because they could be reassured that their queer-bodied children would grow up to be straight-acting adults. But some people didn't like this system: people who--like David Reimer--felt mistreated at best, and medically assaulted at worst, by their medical treatment. Nevertheless, most stayed silent, believing they were alone in their experiences. That changed in 1993, when feminist biologist Anne Fausto-Sterling published articles in *The Sciences* and *The New York Times* exposing the basic fact that intersex exists. In response, "Cheryl Chase wrote a letter" to *The Sciences* announcing the founding of the Intersex Society of North America (ISNA). She founded the group because of her own attempts to recover her history of sex-reassignment in infancy and medically-induced shame, and because of the disinterest of most of her former care providers in what had happened to her. Soon Chase had brought together dozens of people with intersex. Though ISNA began as a support group, it quickly turned into an advocacy group because its members realized that "they had suffered" from similar problems. Like many of the early ISNA members, Chase drew on her political consciousness as a lesbian woman to recognise the degree to which intersex had been unnecessarily socially and medically pathologised. With the successes of the women's health movement and the queer rights movements as a backdrop, "people with intersex began agitating"

for openness and reform. Early on, very few medical professionals recognised ISNA's critiques as legitimate. Many responded that the standard of care was necessary, successful, and justified, even going so far as to actively defend lying to patients about their medical histories. Those at the top simply tried to ignore ISNA. As the leader of the newly formed intersex rights movement, Chase moved rapidly, sometimes able to engage in dialog, and having group protests when doctors would not listen. With her professional background in computer science, she was particularly adept at using the tools of the Internet to spread ISNA's message. ISNA also supported the inquiries of researchers like "Suzanne Kessler":/lessonsfromtheintersexed, "Anne Fausto-Sterling":/books/sexing\_the\_body, and "Alice Dreger":/books/ageofethics, and the organisation engaged in media outreach as much as possible. By about 2001, it had become clear to all that the intersex rights movement was not going away. The claims of activists were illustrated in story after story of problematic intersex treatment, as well as in research that strongly suggested gender identity is "not simply a matter of nurture":/node/564. The fact that medical professionals were unable to produce an intersex patient satisfied with his/her childhood treatment negated the claims that the advocacy groups solely represented the experiences of a disgruntled minority. Finally, many medical professionals began to respond to calls for outcome data, research, full disclosure of information, and revision of homophobic and sexist protocols. The intersex rights movement undoubtedly was helped in its success by surrounding trends in favour of LGBT (lesbian, bi, gay, transgender) rights, patients' rights, and children's rights. Since 1993, due to increased public education, tens of millions of people have learned about intersex. Thanks to the internet, thousands of people with intersex have met others like them, in spite of having been told by their doctors they would never be able to do that--their conditions were supposedly "so rare". ISNA members have gone from picket lines to having a seat at the table in medical conferences. We give grand rounds presentations, help with medical school curricular development, and receive emails from physicians asking for our advice on how to handle intersex cases. Our website is recognized as the definitive source for all things intersex, and for being a life-saving porthold for thousands of people desperate for answers and directions to "their tribe". We have convinced hospitals around the world to examine their practices, to find out what has happened to former patients, and to be accountable for the sometimes-poor effects of good intentions. Over a decade into the work of ISNA, medical professionals are less inclined to lie to patients and parents in intersex cases, are less likely to make openly homophobic or sexist remarks, and are more likely to admit uncertainty about the right course of action. A number of teams are engaged in active outcomes research, though opinions still differ about what outcomes should be sought; some think stable gender identity and heterosexuality are the objective; others suggest it should be lack of depression.) What type of care an individual or family will receive now varies dramatically; what happens to a child with intersex today appears to depend not only on where she or he is born, but who happens to be on call when she or he is born. But we're not done. Even today, the goal of many leading teams treating intersex is still to "make intersex disappear". Paediatric endocrinologist Maria New, recommends Dexamethasone to women who may be carrying an XX child with CAH; these treatments do not alleviate CAH, it only makes the child's clitoris appear smaller (and, clinicians hope, makes the child less likely to grow up lesbian). Abortion is routinely offered to women who are likely pregnant with children with intersex conditions, including Klinefelter's Syndrome. Many surgeons maintain the paternalistic attitude that they should remove healthy testes from babies with AIS to "spare them the trauma later," thereby denying these girls the opportunity to have a natural puberty and to come to know themselves, in a sexual way, free

from surgical scars. Many endocrinologists press unnecessary--sometimes devastating--"normalising" hormone treatments on patients who are otherwise healthy. Finally, doctors continue constructing vaginas in infants and young children, despite arguments by many medical professionals that early vaginoplasties fail too often and are unnecessary to begin with. By contrast, as in the women's rights movement, the civil rights movement, and the LGBT rights movements, the goal of intersex advocacy groups is to have people understand intersex conditions as human rights issues. ISNA maintains as its fundamental principle the principle also fundamental to the women's health movement and the LGBT rights movements: that one's genitals are primarily for one's own use, not for the comfort of others.

[www.isna.org](http://www.isna.org) Intersex Society of North America