[Template:Intersex sidebar](/wiki/Template:Intersex_sidebar" \o "Template:Intersex sidebar) [thumb|300px|Participants at the third](/wiki/File:Third_International_Intersex_Forum.jpg) [International Intersex Forum](/wiki/International_Intersex_Forum), [Malta](/wiki/Malta), in December 2013 **Intersex**, in humans and other animals, describes variations in [sex](/wiki/Sex) characteristics including [chromosomes](/wiki/Chromosome), [gonads](/wiki/Gonad), [sex hormones](/wiki/Sex_hormones), or [genitals](/wiki/Genital) that, according to the UN [Office of the High Commissioner for Human Rights](/wiki/Office_of_the_High_Commissioner_for_Human_Rights), "do not fit typical binary notions of [male](/wiki/Male) or [female](/wiki/Female) bodies".[[1]](#cite_note-1) Such variations may involve genital ambiguity, and combinations of chromosomal [genotype](/wiki/Genotype) and sexual [phenotype](/wiki/Phenotype) other than [XY-male and XX-female](/wiki/XY_sex-determination_system).[[2]](#cite_note-2)[[3]](#cite_note-3) Intersex people were previously referred to as [hermaphrodites](/wiki/Hermaphrodite), but the term has fallen out of favor as it is considered to be misleading and stigmatizing.[Template:Citation needed](/wiki/Template:Citation_needed) Medical description of intersex traits as [disorders of sex development](/wiki/Disorders_of_sex_development) has been controversial[[4]](#cite_note-4)[[5]](#cite_note-5)[[6]](#cite_note-6) since the label was introduced in 2006.[[7]](#cite_note-7) Some intersex infants and children, such as those with ambiguous outer genitalia, are surgically or hormonally altered to create more socially acceptable sex characteristics. However, this is considered controversial, with no firm evidence of good outcomes.[[8]](#cite_note-8) Such treatments may involve sterilization. Adults, including elite female athletes, have also been subjects of such treatment.[[9]](#cite_note-9)<ref name=Chand>[Template:Cite news](/wiki/Template:Cite_news)</ref> Increasingly these issues are recognized as [human rights](/wiki/Intersex_human_rights) abuses, with statements from international[[10]](#cite_note-10)[[11]](#cite_note-11) and national human rights and ethics institutions.[[12]](#cite_note-12)[[13]](#cite_note-13) Intersex organizations have also issued statements, including joint statements as part of an [International Intersex Forum](/wiki/International_Intersex_Forum).

In 2011, [Christiane Völling](/wiki/Christiane_Völling) became the first intersex person known to have successfully sued for damages in a case brought for non-consensual surgical intervention.[[14]](#cite_note-14) In April 2015, [Malta](/wiki/Malta) became the first country to outlaw non-consensual medical interventions to modify sex anatomy, including that of intersex people.[[15]](#cite_note-15)[[16]](#cite_note-16) Like all individuals, intersex people have various [gender identities](/wiki/Gender_identities). Some intersex individuals may be raised as a woman or man but then identify with another gender identity later in life, while most do not.[[2]](#cite_note-2)[[3]](#cite_note-3)[[17]](#cite_note-17)[[18]](#cite_note-18) Most identify as either a woman or man, while some may identify as [neither exclusively a woman nor exclusively a man](/wiki/Non-binary).

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## Definitions[[edit](/index.php?title=(none)&action=edit&section=1)]

According to the UN [Office of the High Commissioner for Human Rights](/wiki/Office_of_the_High_Commissioner_for_Human_Rights):

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In biological terms, [sex](/wiki/Biological_sex) may be determined by five factors present at birth:[[19]](#cite_note-19)

* the number and type of [sex chromosomes](/wiki/Sex_chromosome);
* the type of [gonads](/wiki/Gonad)—ovaries or testicles;
* the [sex hormones](/wiki/Sex_hormone);
* the internal reproductive anatomy (such as the [uterus](/wiki/Uterus) in females); and
* the external genitalia.

People whose five characteristics are not either all typically male or all typically female at birth are intersex.[[20]](#cite_note-20) Some intersex traits are not always visible at birth; some babies may be born with ambiguous genitals, while others may have ambiguous internal organs (testes and ovaries). Others will not become aware that they are intersex unless they receive genetic testing, because it does not manifest in their phenotype.

## History[[edit](/index.php?title=(none)&action=edit&section=2)]

Intersex people are treated in different ways by different cultures. In most societies, intersex people have been expected to conform to either a female or a male [gender role](/wiki/Gender_role).[[21]](#cite_note-21) Whether or not they were socially tolerated or accepted by any particular culture, the existence of intersex people was known to many ancient and pre-modern cultures. An example is one of the [Sumerian](/wiki/Sumer) [creation myths](/wiki/Creation_myth) from more than 4,000 years ago. The story has [Ninmah](/wiki/Ninmah), a [mother goddess](/wiki/Mother_goddess), fashioning humanity out of clay.[[22]](#cite_note-22) She boasts that she will determine the fate – good or bad – for all she fashions:

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The Greek historian [Diodorus Siculus](/wiki/Diodorus_Siculus) wrote of "hermaphroditus" in the first century BCE:

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Numerous historical accounts exist, including those of [Le Van Duyet](/wiki/Le_Van_Duyet) (18th/19th-century Vietnam), [Thomas(ine) Hall](/wiki/Thomas(ine)_Hall) (17th-century USA), [Gottlieb Göttlich](/wiki/Gottlieb_Göttlich) (19th-century Germany), and [Levi Suydam](/wiki/Levi_Suydam) (19th-century USA). The memoirs of 19th-century Frenchwoman [Herculine Barbin](/wiki/Herculine_Barbin) were published by [Michel Foucault](/wiki/Michel_Foucault) in 1980.

During the [Victorian era](/wiki/Victorian_era), medical authors introduced the terms "true hermaphrodite" for an individual who has both ovarian and testicular tissue, verified under a microscope, "male pseudo-hermaphrodite" for a person with testicular tissue, but either female or ambiguous sexual anatomy, and "female pseudo-hermaphrodite" for a person with ovarian tissue, but either male or ambiguous sexual anatomy. In Europe, the term 'intersexual' was first to be used before the Second World War.[[23]](#cite_note-23)[[24]](#cite_note-24)[[82]](#cite_note-82)<ref name=Wiesemann>[Template:Cite journal](/wiki/Template:Cite_journal)</ref> Clinician and researcher [Milton Diamond](/wiki/Milton_Diamond) stresses the importance of care in the selection of language related to intersex people:

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### Self-identification with the term 'intersex'[[edit](/index.php?title=(none)&action=edit&section=20)]

Some people with intersex traits self-identify as intersex, and some do not.[[83]](#cite_note-83)[[84]](#cite_note-84) Some [intersex organizations](/wiki/List_of_intersex_organizations) reference "intersex people" and "intersex variations or traits"[[85]](#cite_note-85) while others use more medicalized language such as "people with intersex conditions",[[86]](#cite_note-86) or people "with intersex conditions or DSDs (differences of sex development)" and "children born with variations of sex anatomy".[[87]](#cite_note-87)

### Hermaphrodite[[edit](/index.php?title=(none)&action=edit&section=21)]

[Template:Main](/wiki/Template:Main) A [hermaphrodite](/wiki/Hermaphrodite) is a plant or animal that has both male and female reproductive organs. Until the mid-20th century, "hermaphrodite" was used synonymously with "intersex".[[26]](#cite_note-26) Currently, hermaphroditism is not to be confused with intersex, as the former refers only to a specific phenotypical presentation of sex organs and the latter to more complex combination of phenotypical and genotypical presentation. Using "hermaphrodite" to refer to intersex individuals is considered to be stigmatizing and misleading.[[88]](#cite_note-88) In reality, *hermaphrodite* is used for animal and vegetal species in which the possession of both ovaries and testes is either serial or concurrent, and for living organisms without such gonads but present binary form of reproduction, which is part of the typical life history of those species; *intersex* has come to be used when this is not the case.

### Disorders of sex development[[edit](/index.php?title=(none)&action=edit&section=22)]

[Template:Main](/wiki/Template:Main) "[Disorders of sex development](/wiki/Disorders_of_sex_development)" (DSD) is a contested term,<ref name=GDavis>[Template:Cite book](/wiki/Template:Cite_book)</ref>[[5]](#cite_note-5) defined to include congenital conditions in which development of chromosomal, gonadal, or anatomical sex is atypical. Members of the Lawson Wilkins [Pediatric](/wiki/Pediatric) [Endocrine](/wiki/Endocrine) Society and the European Society for Paediatric Endocrinology adopted this term in their "Consensus statement on management of intersex disorders".[[7]](#cite_note-7)[[89]](#cite_note-89) While it adopted the term, to open "many more doors", the now defunct [Intersex Society of North America](/wiki/Intersex_Society_of_North_America) itself remarked that intersex is not a disorder.[[90]](#cite_note-90) Other intersex people, activists, supporters, and academics have contested the adoption of the terminology and its implied status as a "disorder", seeing this as offensive to intersex individuals who do not feel that there is something wrong with them, regard the DSD consensus paper as reinforcing the normativity of early surgical interventions, and criticizing the treatment protocols associated with the new taxonomy.[[91]](#cite_note-91) Alternatives to categorizing intersex conditions as "disorders" have been suggested, including "variations of sex development".[[6]](#cite_note-6) [Organisation Intersex International](/wiki/Organisation_Intersex_International) (OII) questions a disease/disability approach, argues for deferral of intervention unless medically necessary, when fully informed consent of the individual involved is possible, and self-determination of sex/gender orientation and identity.[[92]](#cite_note-92) The UK Intersex Association (UKIA) is also highly critical of the label 'disorders' and points to the fact that there was minimal involvement of intersex representatives in the debate which led to the change in terminology.[[93]](#cite_note-93) In May 2016, [Interact Advocates for Intersex Youth](/wiki/Interact_Advocates_for_Intersex_Youth) published a statement opposing pathologizing language to describe people born with intersex traits, recognizing "increasing general understanding and acceptance of the term "intersex"".[[94]](#cite_note-94)

### LGBT and LGBTI[[edit](/index.php?title=(none)&action=edit&section=23)]

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The relationship of intersex to lesbian, gay, bisexual and trans, and queer communities is complex,[[95]](#cite_note-95) but intersex people are often added to [LGBT](/wiki/LGBT) to create an LGBTI community. Emi Koyama describes how inclusion of intersex in LGBTI can fail to address intersex-specific human rights issues, including creating false impressions "that intersex people's rights are protected" by laws protecting LGBT people, and failing to acknowledge that many intersex people are not LGBT.[[96]](#cite_note-96) [Organisation Intersex International Australia](/wiki/Organisation_Intersex_International_Australia) states that some intersex individuals are same sex attracted, and some are heterosexual, but "LGBTI activism has fought for the rights of people who fall outside of expected binary sex and gender norms."[[97]](#cite_note-97)[[98]](#cite_note-98) Intersex is contrasted with and separate from [*transsexuality*](/wiki/Transsexual),[[99]](#cite_note-99)[[100]](#cite_note-100)[[101]](#cite_note-101) which describes the situation in which a person's [gender identity](/wiki/Gender_identity) does not match the person's assigned sex. It is also contrasted with the broader category of [transgender](/wiki/Transgender) people, which includes transsexual people as well as those whose [gender expression](/wiki/Gender_expression) and behavior do not match societal expectations for their assigned sexes. Some people are both intersex and trans.

Intersex can also be contrasted with [*homosexuality*](/wiki/Homosexuality) or *same-sex attraction*. Numerous studies have shown higher rates of same sex attraction in intersex people,[[102]](#cite_note-102)[[103]](#cite_note-103)[[104]](#cite_note-104) thus research on intersex subjects has been used to explore means of preventing homosexuality.[[102]](#cite_note-102)[[103]](#cite_note-103) In an analysis of the use of [preimplantation genetic diagnosis](/wiki/Preimplantation_genetic_diagnosis) to eliminate intersex traits, Behrmann and Ravitsky find social concepts of sex, gender and sexual orientation to be "intertwined on many levels. Parental choice against intersex may thus conceal biases against same-sex attractedness and gender nonconformity."[[105]](#cite_note-105)

## Population figures[[edit](/index.php?title=(none)&action=edit&section=24)]

The number of intersex people depends on the definition used. While human rights institutions have called for the demedicalisation of intersex traits, as far as possible,[[12]](#cite_note-12)[[33]](#cite_note-33)[[106]](#cite_note-106)[[107]](#cite_note-107) medical definitions are still used at present. The now-defunct [Intersex Society of North America](/wiki/Intersex_Society_of_North_America) stated that:

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Specialist medical attention, may include surgery to assign infants to a given sex category (i.e., male or female).[[30]](#cite_note-30) This is controversial due to the human rights implications.[[33]](#cite_note-33)[[107]](#cite_note-107) According to Blackless, Fausto-Sterling et al., on the other hand, 1.7 percent of human births are intersex.[[108]](#cite_note-108)[[30]](#cite_note-30) According to Leonard Sax intersex should be "restricted to those conditions in which chromosomal sex is inconsistent with phenotypic sex, or in which the phenotype is not classifiable as either male or female", around 0.018%. This definition excludes [Klinefelter syndrome](/wiki/Klinefelter_syndrome) and many other variations.[[109]](#cite_note-109) Given that many conditions excluded from Sax's analysis are termed [disorders of sex development](/wiki/Disorders_of_sex_development), such individuals may be subjected to sex "normalizing" interventions, and so they meet current definitions of intersex in use by UN and other bodies, the statistical analyses by Blackless and Fausto-Sterling have become widely quoted,[[33]](#cite_note-33)[[110]](#cite_note-110) including by clinicians.[[111]](#cite_note-111) The following summarizes those frequency statistics:

|  |  |
| --- | --- |
| **Sex Variation** | **Frequency** |
| Not XX, XY, Klinefelter, or Turner | one in 1,666 births |
| [Klinefelter syndrome](/wiki/Klinefelter_syndrome) (XXY) | one in 1,000 births |
| [Turner syndrome](/wiki/Turner_syndrome) (45,X) | one in 2,710 births[[112]](#cite_note-112) |
| [Androgen insensitivity syndrome](/wiki/Androgen_insensitivity_syndrome) | one in 13,000 births |
| [Partial androgen insensitivity syndrome](/wiki/Partial_androgen_insensitivity_syndrome) | one in 130,000 births |
| Classical [congenital adrenal hyperplasia](/wiki/Congenital_adrenal_hyperplasia) | one in 13,000 births |
| [Late onset adrenal hyperplasia](/wiki/Congenital_adrenal_hyperplasia) | one in 1,000 birth.[[113]](#cite_note-113) |
| [Vaginal agenesis](/wiki/Vaginal_agenesis) | one in 6,000 births |
| [Ovotestes](/wiki/Ovotestes) | one in 83,000 births |
| Idiopathic (no discernable medical cause) | one in 110,000 births |
| Iatrogenic (caused by medical treatment, e.g. progestin administered to pregnant mother) | No estimate |
| [5-alpha-reductase deficiency](/wiki/5-alpha-reductase_deficiency) | No estimate |
| [Mixed gonadal dysgenesis](/wiki/Mixed_gonadal_dysgenesis) | No estimate |
| Complete [gonadal dysgenesis](/wiki/Gonadal_dysgenesis) | one in 150,000 births |
| [Hypospadias](/wiki/Hypospadias) (urethral opening in perineum or along penile shaft) | one in 2,000 births |
| [Epispadias](/wiki/Epispadias) (urethral opening between corona and tip of glans penis) | one in 117,000 births[[114]](#cite_note-114) |

## Medical classifications[[edit](/index.php?title=(none)&action=edit&section=25)]

### Signs[[edit](/index.php?title=(none)&action=edit&section=26)]

#### Ambiguous genitalia[[edit](/index.php?title=(none)&action=edit&section=27)]

[thumb|"Hermaphrodite", 1860, by](/wiki/File:Hermaphrodite_by_Nadar_1.jpg) [Nadar](/wiki/Nadar_(photographer)) Ambiguous genitalia may appear as a large clitoris or as a small penis. [thumb|The](/wiki/File:Quigley_scale_for_androgen_insensitivity_syndrome.jpg) [Quigley scale](/wiki/Quigley_scale) is a method for describing genital development in AIS. Because there is variation in all of the processes of the [development of the sex organs](/wiki/Sex_organ#Development), a child can be born with a [sexual anatomy](/wiki/Sexual_anatomy) that is typically female or feminine in appearance with a larger-than-average [clitoris](/wiki/Clitoris) ([clitoral hypertrophy](/wiki/Clitoral_hypertrophy)) or typically male or masculine in appearance with a smaller-than-average [penis](/wiki/Penis) that is open along the underside. The appearance may be quite ambiguous, describable as female genitals with a very large clitoris and partially fused labia, or as male genitals with a very small penis, completely open along the midline ("[hypospadic](/wiki/Hypospadic)"), and empty scrotum.

Fertility is variable. According to some,[[115]](#cite_note-115)[[116]](#cite_note-116) the distinctions "male pseudohermaphrodite", "female pseudohermaphrodite" and especially "true hermaphrodite"[[117]](#cite_note-117) and [Quigley scale](/wiki/Quigley_scale) are visual rating systems that measure genital virilization or femilization.

#### Other signs[[edit](/index.php?title=(none)&action=edit&section=31)]

In order to help in classification, methods other than a genitalia inspection can be performed. For instance, a [karyotype](/wiki/Karyotype) display of a tissue sample may determine which of the causes of intersex is prevalent in the case.

### Causes[[edit](/index.php?title=(none)&action=edit&section=32)]

[thumb|170px|Adult (38), Klinefelter's 46,XY/47,XXY mosaic diagnosis (19):](/wiki/File:Bodymorphproj_mkg_modA001_20070407_posA06.jpg) [gynecomastia](/wiki/Gynecomastia) [Template:Main](/wiki/Template:Main) The common pathway of [sexual differentiation](/wiki/Sexual_differentiation), where a productive human female has an XX chromosome pair, and a productive male has an XY pair, is relevant to the development of intersex conditions.

During fertilization, the sperm adds either an X (female) or a Y (male) chromosome to the X in the ovum. This determines the genetic sex of the embryo.[[123]](#cite_note-123) During the first weeks of development, genetic male and female fetuses are "anatomically indistinguishable", with primitive gonads beginning to develop during approximately the sixth week of gestation. The gonads, in a "bipotential state", may develop into either testes (the male gonads) or ovaries (the female gonads), depending on the consequent events.[[123]](#cite_note-123) Through the seventh week, genetically female and genetically male fetuses appear identical.

At around eight weeks of gestation, the gonads of an XY embryo differentiate into functional testes, secreting testosterone. Ovarian differentiation, for XX embryos, does not occur until approximately Week 12 of gestation. In normal female differentiation, the [Müllerian duct system](/wiki/Müllerian_duct) develops into the [uterus](/wiki/Uterus), [Fallopian tubes](/wiki/Fallopian_tubes), and inner third of the vagina. In males, the Müllerian duct-inhibiting hormone [MIH](/wiki/Anti-Müllerian_hormone) causes this duct system to regress. Next, androgens cause the development of the [Wolffian duct system](/wiki/Wolffian_duct), which develops into the [vas deferens](/wiki/Vas_deferens), seminal vesicles, and ejaculatory ducts.[[123]](#cite_note-123)By birth, the typical fetus has been completely "sexed" male or female, meaning that the genetic sex (XY-male or XX-female) corresponds with the phenotypical sex; that is to say, genetic sex corresponds with internal and external gonads, and external appearance of the genitals.

### Conditions[[edit](/index.php?title=(none)&action=edit&section=33)]

[Template:Further](/wiki/Template:Further) There are a variety of opinions on what conditions or traits are and are not intersex, dependent on the definition of intersex that is used. Current human rights based definitions stress a broad diversity of sex characteristics that differ from expectations for male or female bodies.[[1]](#cite_note-1) During 2015, the [Council of Europe](/wiki/Council_of_Europe),[[33]](#cite_note-33) the [European Union Agency for Fundamental Rights](/wiki/Fundamental_Rights_Agency)[[106]](#cite_note-106) and [Inter-American Commission on Human Rights](/wiki/Inter-American_Commission_on_Human_Rights)[[107]](#cite_note-107) have called for a review of medical classifications that unnecessarily medicalize intersex traits[[33]](#cite_note-33)[[106]](#cite_note-106)[[107]](#cite_note-107)

|  |  |  |
| --- | --- | --- |
| **X/Y** | **Name** | **Description** |
| XX | [Congenital adrenal hyperplasia](/wiki/Congenital_adrenal_hyperplasia) (CAH) | The most common cause of sexual ambiguity is congenital adrenal hyperplasia (CAH), an endocrine disorder in which the [adrenal glands](/wiki/Adrenal_gland) produce abnormally high levels of virilizing hormones in utero. The genes that cause CAH can now be detected in the developing embryo. As Fausto-Sterling mentioned in chapter 3 of *Sexing the Body*, "a woman who suspects she may be pregnant with a CAH baby (if she or someone in her family carries CAH) can undergo treatment and then get tested." To prevent an XX-CAH child's genitalia from becoming masculinized, a treatment, which includes the use of the steroid dexamethasone, must begin as early as four weeks after formation. Although many do not favor this process because "the safety of this experimental therapy has not been established in rigorously controlled trials", it does allow physicians to detect abnormalities, therefore starting treatment right after birth. Starting treatment as soon as an XX-CAH baby is born not only minimizes, but also may even eliminate the chances of genital surgery from being performed.[[30]](#cite_note-30) In XX-females, this can range from partial masculinization that produces a large clitoris, to virilization and male appearance. The latter applies in particular to [Congenital adrenal hyperplasia due to 21-hydroxylase deficiency](/wiki/Congenital_adrenal_hyperplasia_due_to_21-hydroxylase_deficiency), which is the most common form of CAH.  Individuals born with XX chromosomes affected by [17α-hydroxylase deficiency](/wiki/Congenital_adrenal_hyperplasia_due_to_17_alpha-hydroxylase_deficiency) are born with female internal and external anatomy, but, at puberty, neither the adrenals nor the ovaries can produce sex-hormones, inhibiting breast development and the growth of pubic hair.  See below for XY CAH 17α-hydroxylase deficiency. |
| XX | [Progestin-induced virilisation](/wiki/Progestin-induced_virilisation) | In this case, the excess androgen hormones are caused by use of [progestin](/wiki/Progestin), a drug that was used in the 1950s and 1960s to prevent miscarriage. These individuals normally have internal and external female anatomy, with functional ovaries and will therefore have menstruation. They develop, however, some male secondary sex characteristics and they frequently have unusually large clitorises. In very advanced cases, such children have initially been identified as males.<ref name=hplace>[Template:Cite web](/wiki/Template:Cite_web)</ref> |
| XX | [Freemartinism](/wiki/Freemartin) | This condition occurs commonly in all species of [cattle](/wiki/Bovinae) and affects most females born as a twin to a male. It is rare or unknown in other mammals, including humans. In cattle, the [placentae](/wiki/Placenta) of [fraternal twins](/wiki/Fraternal_twins) usually fuse at some time during the pregnancy, and the twins then share their blood supply. If the twins are of different sexes, male hormones produced in the body of the fetal bull find their way into the body of the fetal heifer (female), and masculinize her. Her sexual organs do not develop fully, and her ovaries may even contain testicular tissue. When adult, such a *freemartin* is very like a normal female in external appearance, but she is infertile, and behaves more like a [castrated](/wiki/Castration) male (a steer). The male twin is not significantly affected, although (if he remains entire) his testes may be slightly reduced in size. The degree of masculinization of the freemartin depends on the stage of pregnancy at which the placental fusion occurs[Template:Spaced ndashin](/wiki/Template:Spaced_ndash) about ten percent of such births no fusion occurs and both calves develop normally as in other mammals. |
| XY | [Androgen insensitivity syndrome](/wiki/Androgen_insensitivity_syndrome) (AIS) | People with AIS have a Y chromosome, (typically XY), but are unable to metabolize androgens in varying degrees.  Cases with typically female appearance and genitalia are said to have complete androgen insensitivity syndrome (CAIS). People with CAIS have a [vagina](/wiki/Vagina) and no [uterus](/wiki/Uterus), [cervix](/wiki/Cervix), or [ovaries](/wiki/Ovaries), and are infertile. The vagina may be shorter than usual, and, in some cases, is nearly absent. Instead of female internal reproductive organs, a person with CAIS has undescended or partially descended testes, of which the person may not even be aware.  In mild and partial androgen insensitivity syndrome (MAIS and PAIS), the body is partially receptive to androgens, so there is virilization to varying degrees. PAIS can result in genital ambiguity, due to limited metabolization of the androgens produced by the testes. Ambiguous genitalia may present as a large clitoris, known as [clitoromegaly](/wiki/Clitoromegaly), or a small penis, which is called [micropenis](/wiki/Micropenis) or microphallus; hypospadias and [cryptorchidism](/wiki/Cryptorchidism) may also be present, with one or both testes undescended, and hypospadias appearing just below the glans on an otherwise typical male penis, or at the base of the shaft, or at the perineum and including a bifid (or cleft) scrotum. |
| XY | [5-alpha-reductase deficiency](/wiki/5-alpha-reductase_deficiency) (5-ARD) | The condition affects individuals with a [Y chromosome](/wiki/Y_chromosome), making their bodies unable to convert testosterone to dihydrotestosterone (DHT). DHT is necessary for the development of male genitalia in utero, and plays no role in female development, so its absence tends to result in ambiguous genitalia at birth; the effects can range from infertility with male genitalia to male underdevelopment with hypospadias to female genitalia with mild clitoromegaly. The frequency is unknown, and children are sometimes misdiagnosed as having AIS.[[124]](#cite_note-124) Individuals can have testes, as well as vagina and labia, and a small penis capable of ejaculation that looks like a clitoris at birth. Such individuals are usually raised as girls. The lack of DHT also limits the development of facial hair. |
| XY | [Congenital adrenal hyperplasia](/wiki/Congenital_adrenal_hyperplasia) (CAH) | In individuals with a Y chromosome (typically XY) who have [Congenital adrenal hyperplasia due to 17 alpha-hydroxylase deficiency](/wiki/Congenital_adrenal_hyperplasia_due_to_17_alpha-hydroxylase_deficiency), CAH inhibits virilization, unlike cases without a Y chromosome. |
| XY | [Persistent Müllerian duct syndrome](/wiki/Persistent_Müllerian_duct_syndrome) (PMDS) | The child has XY chromosomes typical of a male. The child has a male body and an internal uterus and fallopian tubes because his body did not produce [Müllerian inhibiting factor](/wiki/Müllerian_inhibiting_factor) during fetal development. |
| XY | [Anorchia](/wiki/Anorchia) | Individuals with XY chromosomes whose gonads were lost after 14 weeks of fetal development. People with Anorchia have no ability to produce the hormones responsible for developing male secondary sex characteristics nor the means to produce [gametes](/wiki/Gamete) necessary for reproduction due to the lack of gonads. They may develop typically feminine secondary sex characteristics without or despite the administration of androgens to artificially initiate physical sex differentiation (typically planned around the age of puberty). Psychological and neurological gender identity may solidify before the administration of androgens, leading to [gender dysphoria](/wiki/Gender_dysphoria), as anorchic individuals are typically [assigned male at birth](/wiki/Sex_assignment). |
| XY | Gonadal Dysgenesis | It has various causes and are not all genetic; a catch-all category.  It refers to individuals (mostly XY) whose gonads don't develop properly. Clinical features are heterogeneous.[[30]](#cite_note-30) |
| XY | [Hypospadias](/wiki/Hypospadias) | It is caused by various causes,including alterations in testosterone metabolism. The urethra does not run to the tip of the penis. In mild forms, the opening is just shy of the tip; in moderate forms, it is along the shaft; and in severe forms, it may open at the base of the penis.[[125]](#cite_note-125) |
| Other | Unusual chromosomal sex | In addition to the most common XX and XY chromosomal sexes, there are several other possible combinations, for example [Turner syndrome](/wiki/Turner_syndrome) ([XO](/wiki/Turner_syndrome)), [Triple X syndrome](/wiki/Triple_X_syndrome) (XXX), [Klinefelter syndrome](/wiki/Klinefelter_syndrome), (XXY) and variants ([XXYY](/wiki/XXYY_syndrome), [XXXY](/wiki/XXXY_syndrome), [XXXXY](/wiki/49,_XXXXY_syndrome)), [XYY syndrome](/wiki/XYY_syndrome), de la Chapelle syndrome ([XX male](/wiki/XX_male_syndrome)), [Swyer syndrome](/wiki/Swyer_syndrome) (XY female). |
| Other | [Mosaicism](/wiki/Mosaicism) and [chimerism](/wiki/Chimerism) | A mix can occur, where some of the cells of the body have the common XX or XY, while some have one of the less usual chromosomal contents above. Such a mixture is caused by either [mosaicism](/wiki/Mosaicism) or [chimerism](/wiki/Chimerism). In mosaicism, the mixture is caused by a mutation in one of the cells of the embryo after fertilization, whereas chimerism is a fusion of two embryos.  In alternative fashion, it is simply a mixture between XX and XY, and does not have to involve any less-common genotypes in individual cells. This, too, can occur both as chimerism and as a result of one sex chromosome having mutated into the other.[[126]](#cite_note-126) Mosaicism and chimerism may involve chromosomes other than the sex chromosomes, and not result in intersex traits. |

## Medical treatment[[edit](/index.php?title=(none)&action=edit&section=34)]

[Template:Further](/wiki/Template:Further)

### Psychosocial support[[edit](/index.php?title=(none)&action=edit&section=35)]

A 2006 clinician "Consensus Statement on Intersex Disorders and Their Management" attempted to prioritise psychosocial support for children and families, but it also supports surgical intervention with psychosocial rationales such as "minimizing family concern and distress" and "mitigating the risks of stigmatization and gender-identity confusion".[[7]](#cite_note-7) In 2012, the Swiss National Advisory Commission on Biomedical Ethics argued strongly in favour of improved psychosocial support, saying:[[13]](#cite_note-13) [Template:Quotation](/wiki/Template:Quotation)

A joint international statement by [intersex community organizations](/wiki/International_Intersex_Forum) published in 2013 sought, amongst other demands:

[Template:Quotation](/wiki/Template:Quotation)

### Surgery[[edit](/index.php?title=(none)&action=edit&section=36)]

[Template:Further](/wiki/Template:Further) Surgical procedures depend on diagnosis, and there is often concern as to whether surgery should be performed at all. Typically, surgery is performed shortly after birth. Surgery may be necessary to assist in bowel and bladder functions. However, defenders of the practice argue that it is necessary for individuals to be clearly identified as male or female in order for them to function socially. Psychosocial reasons are often stated.[[7]](#cite_note-7) This is criticised by many human rights institutions, and authors including [Morgan Holmes](/wiki/Morgan_Holmes) and [Alice Dreger](/wiki/Alice_Dreger), who say that surgical treatment is socially motivated and, hence, ethically questionable; without evidence, doctors regularly assume that intersex persons cannot have a clear gender identity. Parents may be advised that without surgery, their child will be stigmatized.<ref name=holmes>[Morgan Holmes](/wiki/Morgan_Holmes) (2002). Rethinking the Meaning and Management of Intersexuality. Sexualities, 159–180."</ref>

Unlike other aesthetic surgical procedures performed on infants, such as corrective surgery for a [cleft lip](/wiki/Cleft_lip) (as opposed to a [cleft palate](/wiki/Cleft_palate)), genital surgery may lead to negative consequences for sexual functioning in later life (such as loss of sensation in the genitals, for example, when a [clitoris](/wiki/Clitoris) deemed too large or penis is reduced/removed), or feelings of freakishness and unacceptability, which may have been avoided without the surgery. Further, since almost all such surgeries are undertaken to fashion female genitalia for the child, it is more difficult for the child to present as male if that child later identifies as or is genetically male. 20-50% of surgical cases result in a loss of sexual sensation (Newman 1991, 1992).

Additionally, parents are not often consulted on the decision-making process when choosing the sex of the child. The Intersex Society of North America stated that "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 about their conditions. A lot of people in our culture also had no interest in hearing that sex doesn't come in two simple flavors."[[127]](#cite_note-127) Opponents maintain that there is no compelling evidence that the presumed social benefits of such "normalizing" surgery outweigh the potential costs.[[128]](#cite_note-128)[[129]](#cite_note-129) Intersex advocates and experts have critiqued the necessity of early interventions, citing individual's experiences of intervention and the lack of follow-up studies showing clear benefits. Specialists at the Intersex Clinic at University College London began to publish evidence in 2001 that indicated the harm that can arise as a result of inappropriate interventions, and advised minimising the use of childhood surgical procedures.[[130]](#cite_note-130)[[131]](#cite_note-131)[[132]](#cite_note-132)[[133]](#cite_note-133)[[134]](#cite_note-134)[[135]](#cite_note-135)[[136]](#cite_note-136)[[137]](#cite_note-137)[[138]](#cite_note-138)[[139]](#cite_note-139) Studies have revealed how surgical intervention has had psychological effects, leading to the impact on well-being and quality of life. Genitoplasty, plastic surgery done on the genitalia, does not ensure a successful psychological outcome for the patient and might require psychological support when the patient is trying to distinguish a gender identity.[[140]](#cite_note-140) Other than the possible negative psychological outcomes, surgeries, like with a [vaginoplasty](/wiki/Vaginoplasty), can have physical outcomes, one common one being scarring, which can be a factor to insensitivity.[[30]](#cite_note-30) Other cases where vaginoplasty has caused complications, is that the implant or artificial vagina will not stay in place, or need further surgeries.[[141]](#cite_note-141) One of the reasons there are many complications is that doctors who do not specialize in genitoplasty or similar surgeries ([phalloplasty](/wiki/Phalloplasty), vaginoplasty) usually reconstruct the child's ambiguous genitalia.

The Swiss National Advisory Commission on Biomedical Ethics describes surgical interventions as problematic, with "harmful consequences may include, for example, loss of fertility and sexual sensitivity, chronic pain, or pain associated with dilation (bougienage) of a surgically created vagina, with traumatizing effects for the child. If such interventions are performed solely with a view to integration of the child into a family and social environment, then they run counter to the child's welfare. In addition, there is no guarantee that the intended purpose (integration) will be achieved."[[13]](#cite_note-13) In 2013, a submission by the Australasian Paediatric Endocrine Association to an [Australian Senate](/wiki/Australian_Senate) inquiry on the *Involuntary and coerced sterilisation of intersex people in Australia*[[12]](#cite_note-12) acknowledged that there is no firm evidence of good outcomes from appearance-related genital surgeries on infants and children. They state there is "particular concern" regarding post-surgical "sexual function and sensation".[[8]](#cite_note-8) In 2015, an editorial in the [British Medical Journal (BMJ)](/wiki/The_BMJ) described current surgical interventions as experimental, stating that clinical confidence in constructing "normal" genital anatomies has not been borne out, and that medically credible pathways other than surgery do not yet exist.[[142]](#cite_note-142)

### Decision-making on cancer risks[[edit](/index.php?title=(none)&action=edit&section=37)]

In the cases where nonfunctional testes are present, there is a risk that these develop cancer. Therefore, doctors either remove them by [orchidectomy](/wiki/Orchidectomy) or monitor them carefully. This is the case for instance in [androgen insensitivity syndrome](/wiki/Androgen_insensitivity_syndrome).[[143]](#cite_note-143) In a major Parliamentary report in Australia, published in October 2013, the Senate Community Affairs References committee was "disturbed" by the possible implications of current practices in the treatment of cancer risk. The committee stated: "clinical intervention pathways stated to be based on probabilities of cancer risk may be encapsulating treatment decisions based on other factors, such as the desire to conduct normalising surgery… Treating cancer may be regarded as unambiguously therapeutic treatment, while normalising surgery may not. Thus basing a decision on cancer risk might avoid the need for court oversight in a way that a decision based on other factors might not. The committee is disturbed by the possible implications of this..."[[12]](#cite_note-12)

### Medical photography[[edit](/index.php?title=(none)&action=edit&section=38)]

Photographs of intersex children's genitalia are circulated in medical communities for documentary purposes; an example appears on this page. Problems associated with experiences of medical photography of intersex children have been discussed<ref name=PREV>[Template:Cite book](/wiki/Template:Cite_book) p. 72.</ref> along with their ethics, control and usage.[[144]](#cite_note-144) "The experience of being photographed has exemplified for many people with intersex conditions the powerlessness and humiliation felt during medical investigations and interventions".<ref name=CRPH>[Template:Cite news](/wiki/Template:Cite_news) p. 70.</ref>

### Hormone treatment[[edit](/index.php?title=(none)&action=edit&section=39)]

There is widespread evidence of prenatal testing and hormone treatment to prevent intersex traits.[[145]](#cite_note-145)[[146]](#cite_note-146) In 1990, a paper by Heino Meyer-Bahlburg titled *Will Prenatal Hormone Treatment Prevent Homosexuality?* was published in the Journal of Child and Adolescent Psychopharmacology. It examined the use of "prenatal hormone screening or treatment for the prevention of homosexuality" using research conducted on foetuses with [congenital adrenal hyperplasia](/wiki/Congenital_adrenal_hyperplasia) (CAH). Dreger, Feder, and [Tamar-Mattis](/wiki/Anne_Tamar-Mattis) describe how later research constructs "low interest in babies and men – and even interest in what they consider to be men's occupations and games – as "abnormal", and potentially preventable with prenatal dex[amethasone]".[[145]](#cite_note-145)

### Genetic selection and terminations[[edit](/index.php?title=(none)&action=edit&section=40)]

The ethics of [preimplantation genetic diagnosis](/wiki/Preimplantation_genetic_diagnosis) to select against intersex traits was the subject of 11 papers in the October 2013 issue of the [*American Journal of Bioethics*](/wiki/American_Journal_of_Bioethics).[[147]](#cite_note-147) There is widespread evidence of [pregnancy terminations](/wiki/Abortion) arising from prenatal testing, as well prenatal hormone treatment to prevent intersex traits.

In April 2014, [Organisation Intersex International Australia](/wiki/Organisation_Intersex_International_Australia) made a submission on genetic selection via [Preimplantation genetic diagnosis](/wiki/Preimplantation_genetic_diagnosis) to the [National Health and Medical Research Council](/wiki/National_Health_and_Medical_Research_Council) recommending that deselection of embryos and foetuses on grounds of intersex status should not be permitted. It quoted research by Professors [Morgan Holmes](/wiki/Morgan_Holmes), Jeff Nisker, associate professor [Georgiann Davis](/wiki/Georgiann_Davis), and by Jason Behrmann and Vardit Ravitsky.[[148]](#cite_note-148) It quotes research showing [pregnancy termination](/wiki/Abortion) rates of up to 88% in [47,XXY](/wiki/Non-Klinefelter_XXY) even while the [World Health Organization](/wiki/World_Health_Organization) describes the trait as "compatible with normal life expectancy", and "often undiagnosed".[[149]](#cite_note-149)[[150]](#cite_note-150) Behrmann and Ravitsky find social concepts of sex, gender and sexual orientation to be "intertwined on many levels. Parental choice against intersex may thus conceal biases against same-sex attractedness and gender nonconformity."[[151]](#cite_note-151)

### Gender dysphoria[[edit](/index.php?title=(none)&action=edit&section=41)]

The [DSM-5](/wiki/DSM-5) included a change from using [Gender Identity Disorder](/wiki/Gender_Identity_Disorder) to *Gender Dysphoria*. This revised code now specifically includes intersex people who do not identify with their sex assigned at birth, using the language of [Disorders of Sex Development](/wiki/Disorders_of_Sex_Development).[[152]](#cite_note-152) This move was criticised by intersex advocacy groups in Australia and New Zealand.[[153]](#cite_note-153)

## Notes[[edit](/index.php?title=(none)&action=edit&section=42)]

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