



Intersex Article 2: Conditions

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The majority of intersex people will have one of the following conditions but there are also some much rarer and less well researched intersex conditions which are not described below.

The majority of births recorded as intersex are cases of hypospadias. This is not considered by everyone to be a 'true' intersex condition because it has only a very localised effect on bodies which are otherwise male. Hypospadias means that the opening of the urethra (the tube which urine and semen pass through) is not at the tip of the penis but is somewhere else along the length of it, sometimes at the base. This does not normally cause medical problems, but some doctors worry that it can lead to social stigmatisation because boys will be unable to urinate standing up in the normal fashion. It is common for children with hypospadias to be operated on to try and make them look more normal. Surgery of this type is notoriously difficult and it is common for such children to require several operations. Complications can leave a significant number of patients unable to urinate at all without mechanical assistance.

The most common intersex condition to affect people with XX chromosomes is congenital adrenal hyperplasia, or CAH. This is a condition which can be passed down in families. It causes the masculinisation of female fetuses, with varying degrees of severity. CAH babies usually have enlarged clitorises which may look more like penises. These do not cause any medical problems, but are sometimes operated on for cosmetic reasons, in the hope that the reduction will make it easier for the child to grow up as an ordinary girl. However, the evidence suggests that only about fifty percent of CAH people go on to identify as female. In the absence of hormone treatment, CAH people usually develop male secondary sexual characteristics (such as facial hair and deep voices) at puberty. CAH also causes medical problems in most cases, and most CAH people have to take thyroid medication throughout their lives.

A common intersex condition affecting people with XY chromosomes is androgen insensitivity syndrome, or AIS. In AIS cases, the developing foetus fails, either partially or completely, to respond to the presence of testosterone

and so does not develop normal male sex characteristics. In fact, many AIS people are assumed to be ordinary girls when they are born, and it's possible that a significant number are never identified as intersex. The vast majority of AIS people (but not all) identify simply as women. They have short vaginas but lack female reproductive systems; instead they have undescended testicles. Sometimes these descend at puberty, often causing great emotional distress. If they do not descend, most doctors advise that they should be removed, as they have a significant chance of becoming cancerous. AIS people are infertile.

AIS people tend to grow tall like men, but develop female secondary sexual characteristics (such as breasts) and a bone structure which is mostly feminine in appearance.

Some people don't have the usual XX or XY chromosome pairings, but have XXY chromosomes instead. This is known as Klinefelter's syndrome. Most people with Klinefelter's have small penises and small testicles, and many develop breasts at puberty. Most (but not all) identify as male. Klinefelter's is also connected with a range of medical problems. It has been linked to language learning disorders, breast cancer, and a tendency to obesity. The life expectancy for people with Klinefelter's is lower than average; however, the evidence suggests that most are still able to lead fulfilling lives. A few are able to father children.

Another condition in which sex chromosomes are unusual is chimerism. This occurs during gestation when two embryos fuse together at an early stage of development, creating a child who has cells from each of two different genetic combinations. There is increasing evidence that chimerism is in fact quite common, but usually it has little effect on development. However, the fusion of male and female embryos can create a person who has some XX and some XY cells. In some cases, this can lead to the development of intersex characteristics.

Although it is comparatively rare, 5-alpha reductase deficiency syndrome is among the better known intersex conditions. This is partly because it is inherited in such a way that it can be tracked across generations, tending to occur in small clusters in isolated populations; and partly because of the startling way in which it manifests. In Papua New Guinea it is known as kwolu-aatmwol ('female thing changing into male'); in the Dominican Republic, as guevedoche ('balls at twelve'). Babies with this condition appear female although they have male chromosomes. This is because their bodies have not responded normally to testosterone. However, at puberty they develop small penises and male secondary sexual characteristics. In places where this condition is common, it is

not treated with the same secrecy as most intersex conditions are in other cultures. Surgical intervention is rare and most affected people live comparatively normal lives.

Intersex physical bodies can also be caused by a number of lesser known conditions. In some cases the cause cannot be clearly established. In other cases, it is caused by exposure to radiation or to particular chemicals during gestation. During the 1950s and 1960s, many American women were treated with a drug called progestin which it was believed would lower their risk of miscarriage. This drug had the effect of masculinising female foetuses, resulting in a condition resembling CAH. Another cluster of intersex births occurred in eastern France in 1986 and 1987. This is thought to be a result of radiation from the Chernobyl disaster, as prevailing winds carried a lot of radioactive clouds over the area in the immediate aftermath of the disaster and government warnings against eating produce which had been exposed to the rain were not issued until several days later. In both these cases, the intersex births were initially hushed-up, but the truth emerged as those affected grew older and began to encounter one another.

Different people have different personal definitions of what constitutes an intersex condition. Some intersex websites and support groups limit themselves to one or more of the most common conditions, sometimes because it is difficult to provide in-depth information about all the different forms of intersex cases and sometimes because the experiences of people with different intersex conditions do not have much in common. In other cases, intersex people with different conditions join together because of shared concerns about how they are treated by wider society.