## What produces the sexual identity of intersexes?

A study of people with ambiguous genitalia gives unusual support to the prime role of the environment and upbringing in shaping human sexuality. The majority of intersexes (people of ambiguous gender appearance) who have come to the attention of researchers have opted for the gender of upbringing rather than their chromosomal gender. Only a small minority would greatly prefer to change.

Sometimes babies are born with such ambiguous genitalia that medical staff do not know whether the child is a boy or a girl. Until about the 1980s, parents instructed to raise these children in one gender or other often found them developing physically (usually with the onset of puberty) contrary to the gender of upbringing. But, when these children were given the option of corrective surgery and hormonal intervention at puberty, $90 \%$ of those whose cases have been researched opted for the gender in which they had been raised, rather than their biological gender, even in the face of quite contrary physical characteristics. In many cases-though not without difficulty-these children grew up to develop gender behaviours consistent with their gender of choice, rather than their biological gender. They felt attraction, experienced erotic arousal, fell in love in ways characteristic of their chosen gender, married, and raised children.

Today diagnosis is much more sophisticated and medical options much wider. The situation varies a little from country to country, showing social conditioning is important. Many more elect to change from female to male than the reverse because they think being male is better, but overall, lumping all intersex conditions together, about $90 \%$ still choose to remain in the gender of upbringing.

## A lesson in biology

Almost everyone, including homosexuals and lesbians is born chromosomally female (XX) or chromosomally male (XY). When a male sperm carrying 23 chromosomes unites with the female ovum, also carrying 23 chromosomes, the fertilized egg quickly becomes a 46 chromosome cell of 23 pairs, one of each pair from the father, one from the mother. All the chromosomes carry the genetic material that gives us our biological characteristics, but the 23 rd pair is the sex chromosomes, usually comprised of one X chromosome inherited from the mother and an X or Y chromosome inherited from the father. An XX combination in the fertilized egg produces a female, and an XY combination produces a male. Sometimes these standard combinations do not happen, and rare combinations result for reasons that are still not very clear to researchers. One of the X chromosomes can be "lost," leaving only a single X. These fertilized X cells still grow normally, but produce individuals who are very short ( $41 / 2-5 \mathrm{ft}: 137 \mathrm{~cm}+$ ) and physically female, but have no ovaries and are infertile, a condition called Turner's syndrome. Some fertilised cells can end up XXX, resulting in women with a normal female body, but diminished fertility, and sometimes mental retardation. Males can be XYY, with male body type, reduced fertility, and increased height; XXY or XXXY (Klinefelter's syndrome) both cause male body type, but with unusually small penises, shrunken testes, and varying but low production of the male hormone, testosterone, so that at puberty they become only moderately masculine and have scant body hair. The percentage of homosexuals among people with Klinefelter's syndrome is about typical for the general population (contrary to a commonly circulating myth), but about half of those with the syndrome have no interest in any type of sex (they are quite prone to sexual anxiety), partly due to their physical attributes.

Such varied sexuality means a person's chromosomal pattern is not forcing any particular sexuality upon them. ${ }^{1}$

There are all sorts of rare combinations of X and Y , but, in general, if people have an XY or XX combination of some kind they will develop physically as male or female respectively. ${ }^{2}$

## Overwhelming effects of rearing?

John Money, Anke Ehrhardt, and John and Joan Hampson, at the Johns Hopkins Medical School in Baltimore, Maryland, spent a lifetime studying unusual sexual conditions and intersexes. What they found in the sixties about the role of upbringing in the formation of gender identity and sexual orientation led them to the conclusion that the influence of upbringing and rearing was so overwhelming that it was as if a new-born child was a blank slate, written upon only by the influence of upbringing and socialization. ${ }^{3}$ This ultimately proved to be too extreme a conclusion.

In a summary of all cases of intersexes that had come to their attention (particularly the work of the Hampsons), Money and Ehrhardt said about $90 \%$ chose to remain in their gender of upbringing in spite of contrary biology; that is, despite some or any of the following: contrary chromosomes, gonads, hormonal sex, internal sexual organs or external genital appearance. ${ }^{3}$ They remark that of that $10 \%$ who changed their gender, almost all of them made a female to male change. Although the $90 \%$ established a gender identity consistent with their sex of choice, they did not do it without "difficulty, embarrassment, and shame". This represented the situation until the end of the seventies.

## Boy raised as a girl

We now consider a well known case which flatly contradicted the assurance of the Hampsons and Money, but turns out to be rather misleading because it happens only in a minority of cases.

One of Money's cases ${ }^{2}$ was a boy, one of normal identical twin boys, biologically male in every respect, who suffered a surgical mishap during circumcision by electrocautery at the age of seven months. His penis was cut off flush with the abdominal wall. After months of agonizing, the parents decided, when the boy was seventeen months old, to raise him as a girl, and doctors performed the first stages of feminising surgery. The child was called Joan, wore girl's clothing and hairstyle, and the parents were regularly counselled how to raise her under the circumstances. The identical twin brother was raised as a boy. John Money touted this as a
perfect case showing the malleability of gender because the reports seemed to show the "girl" was adapting well to the change. Her mother made a special effort to keep her in dresses because she was initially resistant to them and preferred jeans. By the age of four she had a clear preference for dresses over slacks, wore bracelets and hair-ribbons, and took pride in her long hair. But Joan was tomboyish, had a lot of energy, and was often the dominant one in a girls' group; mother tried to teach her to be more ladylike. Further treatment was planned after puberty.

When the girl was about 13 , she was interviewed by the British Broadcasting Corporation (BBC) and three psychiatrists, who concluded her gender identity was insecure. She refused to talk about sex. When asked to draw a human figure she drew a man because "women are too difficult." She complained that men had it good in life and women didn't. She had found it difficult to be accepted in her group of girls because she was not very attractive, and because her rather clumsy gait had gained her the nickname "cavewoman." She thought she would rather like to be a mechanic. The BBC panel thought that the transformation had been rather shaky, perhaps even inadvisable. It seemed the attempt to environmentally over-ride the basic biology was a failure and "Joan" became the subject of a scholarly fight between Money and other researchers who believed he should never have been brought up as a girl. ${ }^{4}$ Some of them took the debacle as evidence that gender identity was so fixed at birth, that efforts to change it were futile. One sex researcher in Hawaii, Milton Diamond, argues for a "prenatal (biological) organization," a "built-in bias with which a person interacts with his environment," but an extraordinary flexibility to adjust to an erroneously imposed gender. ${ }^{6}$

Money was accused of suppressing some of the evidence he had accumulated that adaptation to the new sex was much less than perfect. It all came to a head in early $1997^{4}$ when it was revealed that at the time of the BBC interview the cat was already out of the bag; Joan had found out three years before that she was really a boy. A year before the program she had rejected hormone treatment for feminisation. (No wonder she looked somewhat masculine.) A year after the program she began a two year program
of penis reconstruction and began to call herself John. Eventually he married a woman several years his senior and adopted her children. Sadly, several years later he committed suicide, just as his co-twin had, some years before, so this complicated story may be further complicated by some mental illness.

A confusing picture, but one that shows, nevertheless, that gender is not written into our genes or gonads. It is malleable and responds strongly to environmental signals. Before the "cat was out of the bag," the boy was behaving to a large degree like the girl he was being raised to be. Afterwards he decided to co-operate with his then known genetic biology rather than suppress it medically, and he also began the corresponding psychological gender shift.

This tragic story is well known, and even been the subject of a book, but it is less well known that Bradley et al. (1998) ${ }^{5}$ reported a (non-twin) case in which the same accident happened much earlier in life. The boy, brought up as female, clearly identified as female even after many years, reaching young adulthood, but said her sexual orientation was bisexual. So this story is almost the opposite of the one above-sometimes reassignment can work.

There is a later collection of data like this-by Meyer-Bahlburg et al. ${ }^{7}$ in which they managed to find no less than 7 boys whose early accidents had forced the amputation of their penises. They were raised as female, and $69 \%$ stayed that way. One was not happy in her gender assignment ("gender dysphoria"). Telling the person the medical details of the accident did make a differencehalf of those told the medical facts before puberty decided to change to male.

Rather similarly, ${ }^{7}$ of 16 boys born through a prenatal biological accident without penises, brought up as female, 12 or $75 \%$ chose to remain that way though two were rather unhappy with the assignment. Of another 17 with the same condition brought up as males, all remained that way. This shows a theme already apparent -there is a preference in most societies to remain or become male. The authors ${ }^{7}$ also said that there was no good evidence that the prenatal surge of testosterone masculinises the brain. The authors concluded:

The data do not support a theory of full biological determination of gender identity development by prenatal hormones and/or genetic factors, and one must conclude that gender assignment and the concomitant social factors have a major influence on gender outcome. On the other hand a number of female-raised individuals did change gender to male and others developed a possible gender-dysphoria, which indicates that gender assignment does not dictate outcome either.

The critical word is "dictate." But what is surprising is how successful upbringing in a contradictory gender actually was.

These data show that the case of the twin boy (Joan/John) was not typical, though the media exposure suggested it was. Most will stay in the gender they were brought up in, male or female. But such cases are very rare. What happens on average with the more common conditions? Do intersex people stay in the sex of upbringing? We will try and summarise. First we describe one of the most common.

## Adrenogenital syndrome in females (congenital adrenal hyperplasia)

This condition, which affects female fetuses, is the result of a genetic defect; the adrenal glands do not produce their proper hormone, cortisone. Instead, they release a precursor product, which acts as a male hormone, an androgen. This enters the bloodstream of the female fetus too late to masculinise the internal reproductive system, which is already female, but in time to masculinise the external genitalia. The result is a chromosomal female with a uterus and two ovaries, but anything from a grossly enlarged clitoris resembling a penis with partially fused labia (resembling testicles) to a fully formed penis and empty scrotum. Because people with this condition continue to produce androgen for the rest of their lives, they must also take doses of the antidote, cortisone, to counteract it - in childhood, to stop an excessively masculine puberty which comes 8 to 10 years too early, but also in adulthood. Although the ovaries continue to secrete normal levels
of female hormones, these are overwhelmed by the high amounts of androgen being produced by the adrenal gland.

The percentage of these women who are brought up female but want to change to male varies with country and research group: Byne ${ }^{8} 2-10 \%$ changed, Dessens ${ }^{9} 5 \%$ changed, Long ${ }^{10} 0 \%$ (they became indistinguishable from controls by adulthood), Meyer-Bahlburg ${ }^{11} 9 \%$ changed, Reiner ${ }^{12} 22 \%$ changed from female, Slijper ${ }^{13}$ $13 \%$ changed from female. So, overall, about $10 \%$ of these females with adrenogenital syndrome chose to change,

## Turner's Syndrome (single X chromosome)

We met this condition above. It has an interesting bearing on the subject. Because of their lack of ovaries, or non-functional ovaries, all Turner's Syndrome people take estrogen throughout their lives. Some marry men, and recently some research surveyed how they were functioning sexually. The women were in two groups -those in relationships and those not. Approximately $30 \%$ of the study group were involved in a partner relationship, and this group scored within the average range for heterosexual women on fantasy, arousal, experience, orgasm. The authors concluded they had relatively normal overall sexual function, but the majority of unpartnered women reported very low level sexual functioning. ${ }^{14}$

Since both were on estrogen this hormone seems to be a negligible contributor. The conclusion is that sexual functioning arises overwhelmingly out of the partner relationship.

## Males born without gonads

One more intersex study is enlightening. Szarras-Czapnik et al. ${ }^{15}$ reported on 10 males born without testes. What was their sexual orientation? The males cannot have had the prenatal testosterone surge which is supposed to make the brain masculine so this is an interesting test. The males were all solidly male and with a heterosexual orientation-7/10 had had sex with a woman. This argues
again that at least for males, upbringing is far more important than testosterone.

## Biologically-induced gender change

## ( $5 \alpha$-reductase deficiency)

One special genetic condition seemed initially a possible major exception to the general rule of remaining with the gender of upbringing, and this was the deficiency of an enzyme called $5 \alpha-$ reductase. This deficiency prevents formation of one of the male hormones, dihydrotestosterone, so that the usual prenatal surge of testosterone that differentiates a boy from a girl before birth does not occur* and external genitalia are ambiguous. If the condition is not diagnosed and treated, everyone gets a shock at puberty when the testes become detectable and the body becomes masculine. The researchers studied 38 of these cases in the Dominican Republic, particularly 18 who were "unambiguously raised as girls". ${ }^{16}$ Their findings? At puberty or after, 17 of these children changed to a male gender identity and developed an erotic interest in women. Many became heads of families. The researchers argued that androgens made a "strong and definite contribution to male gender identity."

But the conclusion is not as straightforward as it seems. Critics of the research argue that men had much greater status and prestige in Dominican society, and that together with sudden masculinisation, a choice to be male could be strongly culturally influenced. Certainly the Dominican study seems to stand alone in the strength of its argument for a hormonal basis to gender identity. Another researcher into $5-\alpha$ reductase deficiency drew an opposite conclusion. Gilbert Herdt, the most prominent researcher among the Sambia of the eastern highlands of Papua New Guinea, found five cases of $5 \alpha$-reductase deficiency in his study group. ${ }^{17}$ In this case the individuals were raised as girls but on their sudden masculine development at marriageable age (puberty), were treated as a third sex. Although the Sambia are a strongly misogynist culture, there was no attempt-as in the Dominican Republic-to adopt a

[^0]male gender, because the culture forbade it; the Sambia believed a boy could only become a man through ingestion of male semen in prescribed regular fellatio in childhood. Based on this cultural prohibition on becoming male Herdt argues that gender identity is therefore culture dependent rather than hormone dependent. Herdt also maintained that only 13 in the Dominican Republic study, not 17 , lived unequivocally as men. In an almost identical condition found in the Gaza strip, only $28 \%$ changed gender at puberty. ${ }^{18}$

In the West, of those who have $5 \alpha$-reductase deficiency, only a small percentage elect to change gender at puberty, and they find it difficult. ${ }^{19}$ Ninety percent are content to remain in the gender of upbringing (female), possibly because the perceived rewards of being a Western woman are greater than in other cultures.

## Other changes by intersexes later in rearing

In India ${ }^{20}$, of 74 intersex patients, all but one stayed with the sex of rearing. In Egypt ${ }^{21} 10 \%$ changed. Reiner ${ }^{12}$ in a paper from the West found that of 60 raised as female, $43 \%$ declared themselves female but $53 \%$ changed to male. This was a rather exceptional group in which there was considerable decision to change.

In Bahrain, McCarthy ${ }^{22}$ found all female patients reassigned the male gender accepted that, but no males accepted a change to female!

In Russia, according to Lev-Ran ${ }^{23}$, all adult patients resisted reassignment and wanted to remain the way they had been brought up!

It was noticeable that a Western group with cloacal exstrophy ${ }^{7}$ (in which sexual organs are poorly developed and internal organs such as bladder may protrude) had a large percentage-about $33 \%$ - of people who wanted to change from female to male and more who were unhappy.

Since the congenital adrenal hyperplasia group predominates (among intersexes), the original estimate of those who wished to change up to about 1980 still holds and is around $10 \%$. The influence of upbringing is strong, but less so in the West since the days of the Hampsons.

## Should intersex children be allowed to choose?

In the last few decades a strong minority opinion has formed that intersex children should be allowed the maximum choice and there should be no medical intervention at all. A human rights ordinance in San Francisco ${ }^{24}$ sought to make this mandatory. However traditional medical opinion argues strongly that the intersex condition is devastatingly embarrassing for school children and that intervention is far preferable. Less than $10 \%$ of intersex children later disapprove of the early intervention, and in countries such as Vietnam where corrective surgery is not available, intersex adults lament that it wasn't. ${ }^{25}$ However , one report that studied 38 surgical early interventions found that four were much later at serious risk of Gender Identity Disorder. ${ }^{26}$ It seems the medical specialists can't win: if there is no early surgical intervention, about $10 \%$ want to change later in life; if there is early surgical intervention, a different $10 \%$ will want to change.

## Summary

So if we ask the question, "What produces the sexual identity of intersex children?" we have to answer that upbringing greatly predominates-even in modern society, and more so in a less liberal society.

That is, about $90 \%$ of intersexes on record have elected to continue in the gender in which they were raised, even in the face of strongly contradictory biological and physical characteristics. If the influence of upbringing is so strong that it can over-ride obvious contrary biological predispositions, then it is more powerful than biology in the development of gender identity, at least in most countries. It becomes nonsensical to argue that gender identity in chromosomally normal individuals (like homosexuals) is genetically or biologically enforced. In modern Western society, sexual identity appears to be about $10 \%$ genetic and $90 \%$ environmental. So it is quite plausible that homosexuality is also $10 \%$ genetic and $90 \%$ environmental.

## References

1. Vogt HJ. 1984. Sexual behavior in Klinefelter's syndrome. In Klinefelter's Syndrome, ed. Bandmann HJ, Breit R, Perwein E,163-169 pp. Berlin: Springer-Verlag
2. Money J, Ehrhardt AA. 1972. Man \& Woman, Boy \& Girl. Baltimore: The Johns Hopkins University Press
3. Hampson JL, Hampson JG. 1961. The ontogenesis of sexual behavior in man. In Sex and Internal Secretions, ed. Young WC, Corner GW,1401-1432 pp. Baltimore: Williams and Wilkins.
4. Diamond M. 1982. Sexual identity, monozygotic twins reared in discordant sex roles and a BBC follow-up. Archives of Sexual Behavior 11:181-6
5. Bradley SJ, Oliver GD, Chernick AB, Zucker KJ. 1998. Experiment of nurture: ablatio penis at 2 months, sex reassignment at 7 months, and a psychosexual follow-up in young adulthood. Pediatrics 102(1):e9
6. Diamond M. 1965. A critical review of the ontogeny of human sexual behavior Quarterly Review of Biology 40:147-75
7. Meyer-Bahlburg HFL. 2005. Gender identity outcome in female-raised 46,XY persons with penile agenesis, cloacal exstrophy of the bladder or penile ablation. Archives of Sexual Behavior 34:423-38
8. Byne W. 2006. Developmental Endocrine Influences on Gender Identity: Implications for Management of Disorders of Sex Development. Mt Sinai Journal of Medicine 73:950-9
9. Dessens AB, Slijper FME, Drop SLS. 2005. Gender dysphoria and gender change in chromosomal females with congenital adrenal hyperplasia. Archives of Sexual Behavior 34:389-97
10. Long DN, Wisniewski AB, Migeon CJ. 2004. Gender role across development in adult women with congenital adrenal hyperplasia due to 21-hydroxylase deficiency. Journal of Pediatrics and Endocrinological Metabolism 17:1367-73
11. Meyer-Bahlburg H, Dolezal C, Baker S, Ehrhardt A, New M. 2006. Gender Development in Women with Congenital Adrenal Hyperplasia as a Function of Disorder Severity. Archives of Sexual Behavior 35(6):667-84
12. Reiner WG. 2005. Gender identity and sex-of-rearing in children with disorders of sexual differentiation. Journal of Pediatrics and Endocrinological Metabolism 18:549-53
13. Slijper FME, Drop SLS, Molenaar JC, de Muinck Keizer-Schrama SMPF. 2000. Response to Milton Diamond and Cheryl Chase. Archives of Sexual Behavior 29:119-22
14. Sheaffer AT, Lange E, Bondy CA. 2008. Sexual function in women with Turner syndrome. Journal of Womens Health (Larchmont) 17(1):27-33
15. Szarras-Czapnik M, Lew-Starowicz Z, Zucker KJ. 2007. A psychosexual follow-up study of patients with mixed or partial gonadal dysgenesis. Journal of Pediatric and Adolescent Gynecology 20(6):333-8
16. Imperato-McGinley J, Peterson RE, Gautier T, Sturla E. 1979. Androgens and the evolution of male-gender identity among males pseudohermaphrodites with 5- $\alpha$ alpha-reductase deficiency. The New England Journal of Medicine 300(22):1233-7
17. Herdt G. 1990. Mistaken gender: 5-alpha reductase hermaphroditism and biological reductionism in sexual identity reconsidered. American Anthropologist 92:433-46
18. Rösler A, Kohn G. 1983. Male pseudohermaphroditism due to $17 \beta$ hydroxysteroid dehydrogenase deficiency: studies on the natural history of the defect and effect of androgens on gender role. Journal of Steroid Biochemistry 19:663-74
19. Byne W, Parsons B. 1993. Human sexual orientation. The biologic theories reappraised. Archives of General Psychiatry 50:228-39
20. Ammini AC, Gupta R, Kapoor A, Karak A, Kriplani A, Gupta DK, Kucheria K. 2002. Etiology, clinical profile, gender identity and long-term follow up of patients with ambiguous genitalia in India. Journal of Pediatrics and Endocrinological Metabolism 15:423-30
21. Dessouky NM. 2003. Gender assignment for children with intersex problems: an Egyptian perspective. Saudi Medical Journal 24:S51-S52
22. McCarthy J. 1998. Bahrain. In The International Encyclopedia of Sexuality, ed. Francoeur RT,116-186 pp. New York: Continuum
23. Lev-Ran A. 1974. Gender role differentiation in hermaphrodites. Archives of Sexual Behavior 3:391-423
24. Phillips H. 2001. Boy meets girl. New Scientist 12 May:29-35
25. Dennis C. 2004. The most important sexual organ. Nature 427:390-2
26. Crawford JM, Warne G, Grover S, Southwell BR, Hutson JM. 2009. Results from a pediatric surgical centre justify early intervention in disorders of sex development. Journal of Pediatric Surgery 44(2):413-6

[^0]:    * It is doubtful that the brain is masculinised by the pre-natal testosterone surge but the surge is sufficient to masculinise both internal and external sex organs.

