

CHAPTER 12

Sex Determination

What are little boys made of? The simple answer is: a Y chromosome. In humans and other mammals, females have two large chromosomes, the X chromosomes, and males have one X chromosome and a smaller Y chromosome (which has few or no genes on it other than those determining maleness). Spermatozoa bearing X and Y chromosomes are produced in approximately equal numbers and the sex of the new individual is determined at the time of fertilisation by the type of sperm fertilising the X-bearing egg.

The Y chromosome is extremely powerful. Even individuals with eight X chromosomes and a single Y are male. It was at one time thought that an individual could be male without a Y chromosome but the missing Y was finally found as a tiny piece stuck to an X chromosome.

Not all animals have as straightforward a system as this. In some mammals, females have two copies of each of two different X chromosomes, while males have one copy of each plus a Y chromosome. In others, females have two X but males have an X and two Ys. In some reptiles, sex is determined not by sex chromosomes but by the environment, especially temperature.

The situation in many bony fishes seems even more bizarre to us. These are hermaphrodites that retain the ability to change sex, either under the influence of hormones or because of social factors, in adult life. This appears to be because sex differentiation is triggered, not by a direct effect on the undifferentiated gonads, but by initiating events in the brain. The switch is under genetic influence but is always at least potentially subject to environmental influence.

In mammals, testes and ovaries differentiate from undifferentiated gonads late in development.

The female is the default pattern - i.e., it does not require any specific stimulus. If there is no Y or H-Y antigen present, if an embryo's gonads spontaneously fail to develop or if an embryo's gonads are removed at the sexually undifferentiated stage, the result will be a female.

In the male, the Y chromosome promotes testicular development and carries a gene that codes for, or regulates, the synthesis of a substance called H-Y antigen that is believed to promote testicular differentiation. The foetal testis promotes development of the male secondary sex characteristics and inhibits development of the female structures. It does this by producing two hormones - testosterone, which does the first job, and mullerian-inhibiting hormone, which does the second. If either of these processes is incomplete, hermaphroditism (where both male and female organs are present) or pseudohermaphroditism (where definite male or female organs are present together with less developed structures of the other sex) results. Estimates of the frequency of such intersexuals range from one in 2,000 to as many as four percent of births.

True hermaphrodites are the rarest. There is enormous variation in the exact nature of their sexual equipment, with a slight preponderance of those with a more masculine physique. They also vary considerably in the degree of function of their different structures. Theoretically, it might be possible for a true hermaphrodite to become both father and mother to a child but in practice the appropriate ducts and tubes are never configured so that egg and sperm can meet. Nonetheless, there are rare individuals who are capable of being either father or mother.

Rather more are capable of having normal intercourse with both men and women. Urologist Hugh Young, in his book *Genital Abnormalities, Hermaphroditism and Related Adrenal Disease* (published in 1937), relates the intriguing story of Emma, who had both penis-sized clitoris and vagina. She was brought up as a girl and married at nineteen but had sex with several girls both before and after her marriage (such sex gave her much more pleasure than sex as a woman). When Young told her it would be relatively easy to make her a man, she retorted strongly that she had no wish to lose her meal ticket!

Male pseudohermaphrodites have testes and XY chromosomes but also have a vagina and a clitoris and often develop breasts at puberty; however, they do not menstruate. Female

pseudohermaphrodites have ovaries, two X chromosomes and sometimes a uterus but also have at least partly masculine external genitalia and can develop beards, deep voices and adult-size penises.

The conventional medical wisdom - at least for the last forty years - is that intersexuality is a mistake that should be repaired as soon as possible after birth; i.e., a judgment should be made as to whether the child should be male or female. If a newborn's stretched organ is longer than an inch, he is deemed a he; if the protrusion is under 3/8 inch, she is a she. Those falling in between have their organs shortened, a penis becoming a clitoris. Further plastic surgeries and hormone treatments during puberty complete the transformation, with external female genitalia sculpted from scrotal tissue.

However, because genital plastic surgeries damage sexual function and fail to produce normal genital appearance; because there are a large number of case histories, mostly from between 1930 and 1960, of intersexuals who have adjusted happily and coped with the many admitted problems; and because, as ethicists like Laurence McCullough tirelessly reminds clinicians, intersexuality is a variation and not a disease; there is a strong push for a reappraisal.

There are also several case histories from more recent times where sex assignment against the chromosomal sex has led to great misery for the individual, providing a degree of rebuttal to the extreme feminist viewpoint that sex behaviour is all learned and plastic. Academic work which has been under way for the past five years has resulted in the conclusion that normal male gender identity can develop not only in the absence of the penis but even after the removal of the testicles and unequivocal rearing as female. Male gender identity appears directly related to normal male patterns of hormone exposure in the womb.

On the other hand, people with androgen insensitivity syndrome have male (XY) chromosomes and fully functioning testes but their body cells lack receptors for testosterone. They develop as females and may go through life blissfully unaware that they are biologically male. Those with 5-alpha-reductase deficiency, whose cells possess testosterone receptors but cannot process it, are less fortunate. They start out life looking like girls but, at puberty when their testes start producing much more testosterone, change to boys.

What of those who are biologically of one sex but feel themselves as of the other? Frankly, there is no evidence of any biological basis in these cases and, after their operations, transsexuals biologically remain mutilated males or (rarely) females. However, Dr Harry Benjamin, who changed George to Christine Jorgensen, believed that if you can't change the mind to fit the body, you should change the body to fit the mind. The Harry Benjamin International Gender Dysphoria Association has the stated goal of treatment of 'lasting personal comfort with the gendered self in order to maximise overall psychological well-being and self-fulfillment' - an admirable goal, however achieved.

Human individuals with a pattern of sex chromosomes different from the normal XY one (produced by mistakes in the first cell division after fertilisation) have a number of defects depending on the particular pattern. Those with an X but no Y (Turner's syndrome) develop into females with webbed necks and sexually juvenile development without secondary sex characteristics but have normal intelligence. Individuals with three X chromosomes are generally more or less normal females but those with more than three are usually severely retarded. Those with two (Klinefelter's syndrome) or more X chromosomes and a Y chromosome become sterile males with small testes; some are mentally retarded. On the other hand, those with an X and two Ys (0.1 percent of all males) are taller than usual but otherwise relatively normal, though some show severe aggressive tendencies.

In societies in which inheritance is entirely through one sex (usually male, occasionally female), babies of that sex have always been more highly valued. In many societies, other factors have also operated for both men and women to prefer sons to daughters. The old (and, in some places, not so old) method of achieving this was infanticide. When this became barred by law, other methods were sought. One of the oldest is found in several ancient Greek and Roman texts

and involves various means of taking advantage of the 'fact' that the right side is associated with males and the left with females. Numerous other methods based on observations of supposed correlations followed. When statistics became a science, it was coopted into the search, with no more significant results than to give credence to the persistent myth of the effect of timing.

More recent attempts have been based on studying differences between male (i.e., Y chromosome-bearing) and female (X-bearing) sperm. The technique that claims the most success is one, developed by Ronald Ericsson and available in some clinics, which uses the supposed ability of Y-bearing sperm to swim faster through liquid albumin than X-bearing sperm. A success rate of 77 percent is claimed for this method but these claims have never been independently verified.

At the moment, the only method providing any great assurance of effective sex selection is determination of the sex of the baby early in pregnancy by amniocentesis or chorionic villus sampling and abortion if the foetus is of the 'incorrect' sex. This procedure may be justifiable in cases where either or both parents are carriers of a sex-linked genetic disorder but is also widely performed in India (and probably other places) for other less worthy reasons.

It may soon be possible to use DNA hybridisation to select embryos of a particular sex before they are implanted in women undergoing in vitro fertilisation. The ethical and social implications of such a practice deserve serious discussion. Let us hope - and work towards ensuring - that such judgments are not based purely on an undervaluing of the worth of either sex.