

Being born of uncertain gender is the last sexual taboo. But why is the truth about 'intersex' so often kept from the patients themselves? Christine Toomey reports

THE WORST OF BOTH WORLDS

How would you feel if you'd grown up thinking you were a girl and discovered at 20 that, genetically, you were a man? Or if, as a girl of 18, you found your parents gave you a boy's name when you were born because doctors were confused about your gender? Imagine you are a happily married man of 29, whose only problem is that your wife is failing to conceive, and that tests reveal the problem lies with you - your genes are 100% female. Or imagine you discover your parents had been advised by doctors never to tell you there had been doubts about your gender, or that surgeons operated on you and did not tell you the real reason for the surgery? Would you feel sad, angry, confused?

This is the story of men, women and children who face such problems. They were born with a medical condition called intersex. It is not as unusual as you might suppose, and according to some medical experts is becoming more common.

The large brown envelope that made sense of Melissa's troubled past arrived in the post shortly after her 18th birthday. Melissa took the package to the privacy of her bedroom before breaking the seal on information she hoped would explain the feelings of shame and secrecy that had dominated her childhood and the reasons for the repeated gynaecological surgery she had endured. She was shocked and angered by what she read.

As Melissa was growing up, she had been told that she suffered from a rare metabolic disorder that caused her body to lose excessive amounts of salt and led to her being placed on medication she would need for the rest of her life.

She knew she had been a small, sickly baby who had not been expected to survive. She knew the condition she was born with was called congenital adrenal hyperplasia (CAH). What she was not told, but discovered from the information sent to her that day, was that CAH is an 'intersex' condition. She had been born of mixed gender, with ambiguous-looking genitalia, neither wholly female nor male.

'I was told as I was growing up that something was 'not quite right down there'. But I was also told not to ask too many questions. I thought it must be something horrible,' Melissa, now 33, recalls.

"My mother told me to always use a cubicle, and nobody but a doctor could touch me' "

'My mother just told me that I should always use a cubicle to change at school and that nobody but a doctor was ever allowed to touch me.'

Below Melissa, 33 who was born with congenital adrenal hyperplasia. The truth about her condition was kept from her until 18 years old. 'I was devastated. Not that I was intersex, but that I had been lied to'



When Melissa confronted her mother, and asked her why she had never been told the truth, 'She said she thought the truth would upset me,' says Melissa. 'She told me for the first time that doctors had thought I was a boy when I was born, and that she and my father had named me Nicholas.' But when doctors pumped dye through her genital tract when she was three weeks old, they discovered Melissa had a womb and ovaries. 'My parents were told I was a girl after all, but that I would need surgery to 'normalise' things. I was devastated. Not that I was intersex, but that I had been lied to.'

After requesting that her medical records be released to her, Melissa discovered that the operations she had undergone as a baby, and later at the ages of 4, 12 and 14, consisted of a complete removal of the clitoris - hers was considered too large. She also underwent extensive reconstruction of her vagina. 'The outcome,' she says, 'was a mess.'

Below Melissa aged three

Anna was also never told the truth about her condition, or the real reason she underwent surgery at the age of 20. After seeking advice from her doctor about her failure to menstruate, Anna was sent to a London hospital for examination, where she was told she had lumps in her abdomen that needed to be removed. After surgery, she was prescribed a high dosage of the female hormone oestrogen and discharged with little follow-up.

'I felt confused, like something was very wrong. But I didn't understand what was happening,' says Anna, now 44. 'When I looked through some medical notes that were left by my bed in hospital, I saw someone had written, 'She is not aware of her condition.' When I asked my mother what that meant, she told me never to ask the doctors about it.'

In the years that followed, Anna tried to find out more about what had happened to her in hospital. Her mother gave her 'a few snippets of information to go on', she says, almost casually, such as the fact that she was born with XY chromosomes, which meant that genetically she had been born male, not female. It was then, she says, that her childhood began to make sense. Anna remembers quite clearly at school feeling she did not belong with the other girls. 'I looked like a girl. But I knew I was a boy.



I always played with the boys. As a teenager I fell in love with girls. But I didn't become sexually active.'

What Anna slowly discovered in medical journals was that she had a rare genetic disorder called androgen insensitivity syndrome (AIS), also classed as an intersex condition. This meant that although externally she looked like a girl, she had no womb or ovaries, her vagina was undeveloped, and her body produced high levels of testosterone.

The lumps that doctors removed when she was 20 were undescended testes. 'Because they kept what they were doing secret, I had no way of knowing what a huge impact removing this source of testosterone would have on my psyche, no way of objecting to it,' says Anna.

'If I had been 100% male, they would never have got away with it. But if you are mixed gender you are treated appallingly, as if you have no rights.'

Those born with intersex conditions are often referred to, incorrectly, as hermaphrodites.

True hermaphrodites are comparatively rare, as they are born with gonadal tissue capable of producing both sperm and eggs. Intersex covers a much wider spectrum of medical conditions, including chromosomal abnormalities in which a person's genitals do not match their chromosomes, or do not conform to male or female norms (see panel, right). According to some estimates, as many as 1 in 2,000 babies are born with one of these conditions. If accurate, this means that more babies are born intersex than are born with cystic

fibrosis, the incidence of which is 1 in 2,500. Some doctors prefer a stricter definition of the term 'intersex', so argue that this figure is exaggerated. Others believe it is a conservative estimate, arguing that the condition sometimes goes undetected or unreported because of its delicate nature.

Some medical experts believe the incidence of intersex is on the increase. 'It certainly seems to be more common now than 20 or 30 years ago,' says Professor Ieuan Hughes, an endocrinologist at Cambridge university's paediatrics department, based at Addenbrooke's Hospital.

'Certainly, marginal related conditions are becoming more common, like undescended testes,' says Philip Ransley, a consultant urologist at Great Ormond Street Hospital for Children. Some attribute this to the increase in quantities of female hormone entering the environment because of the contraceptive pill or oestrogen-like substances caused by the degradation of some plastics, though no direct link has so far been proven.

Beyond doubt, however, is the culture of shame that can surround the treatment of those with intersex conditions. Many doctors prefer to draw a veil of secrecy around the treatment they give to people with these conditions, on the grounds that discovering the truth would be too traumatic for the patient. 'It is still regular practice in some units not to tell patients their diagnosis,' says Sarah Creighton, a consultant gynaecologist at University College London Hospitals (UCLH), who specialises in the treatment of intersex patients.

'I see many letters sent from consultants to GPs – particularly concerning the treatment of women - saying, 'She is fine. She does not know her diagnosis and I do not feel it is appropriate for her to learn it, as it would be devastating.'

But growing pressure from patients who believe their lives have been ruined by surgery, performed without their consent, may change such attitudes. Angry patients in many countries have called for a moratorium on early surgery. The only circumstances in which early genital surgery should be performed, they argue, is if a condition leads to medical complications - undescended testes can, for instance, cause hernias, and testicular tissue can, in some intersex patients, become cancerous. Intersex individuals - and a growing number of doctors who treat them - argue that those born with such conditions should be allowed to decide which gender they most identify with when they mature. This has made intersex management one of the most talked-about subjects in medicine and the focus of highly charged debate.

The British Association of Paediatric Surgeons recently set up a working party to look at new guidelines for the treatment of babies born with intersex conditions, and it is expected to advise against early genital surgery. But some physicians are indignant that their medical judgment should be called into question regarding the best treatment for patients. Philip Ransley, for instance, dismisses activists against early surgery in the United States as 'green-wellied loonies'.

Ransley argues that anyone who recommends a blanket policy of 'Don't do anything, wait and see' is failing to grasp the different treatment each condition requires. 'Part of the problem with the debate is the use of sweeping generalisations that do not apply in specific cases,' he says. 'The surgery for most of these conditions changed radically between 10 and 15 years ago; nobody has sufficient information yet to know if those changes have been beneficial.'

Part of the controversy surrounding the treatment of intersex is about what ranks as aesthetically acceptable genitalia. 'Part of this debate is about how we can balance the rights of the individual against the pressures that will be brought to bear on that individual because of their appearance,' says Peter Hindmarsh, a leading endocrinologist also at Great Ormond Street. 'Medicine does not usually accept the views of society if they are in conflict with the needs of the individual.'

HOW SEXUAL DIFFERENCE GETS MIXED UP

Intersex conditions fall into two broad categories: those caused by sex chromosome abnormalities and those termed variants of hermaphroditism. Not all of these conditions are detected at birth; some only become apparent after a girl fails to menstruate or develops a groin hernia, or once a man or woman discovers a problem with fertility. This was poignantly illustrated by the case of a 29-year-old man from East Anglia who, after his wife failed to conceive, was found to be 100% genetically female: blood tests revealed his chromosomes were XX. Devastated, he refused any medical treatment, has refused to be identified and has had no further contact with the hospital that carried out the tests. While not all sex chromosome irregularities lead to abnormal genitalia and reproductive organs, the most common do

KLINFELTER SYNDROME
Caused by an extra X chromosome, people with this condition possess male reproductive organs and genitalia. However, the testes fail to develop at puberty, the penis is often small and sperm is not produced.

TURNER'S SYNDROME
This condition, the result of only one X chromosome being present, often leads to spontaneous abortion of the foetus. Babies who survive develop as females. If untreated, they fail to develop breasts and, because of undeveloped ovaries, are infertile.

CONGENITAL ADRENAL HYPERPLASIA (CAH)
The most common cause of female pseudohermaphroditism, and the condition suffered by Melissa, is caused by a missing enzyme, which causes overstimulation of the adrenal gland, overproduction of the

male hormone androgen, and virilisation of the genitalia. Sufferers have XX chromosomes, ovaries and are genetically female.

ANDROGEN INSENSITIVITY SYNDROME (AIS)
The most common cause of male pseudohermaphroditism, caused by a defective gene that prevents genetically male fetuses (XY) from absorbing androgen. It is the condition from which Anna suffers. The foetus develops testes, but continues to develop along female lines, although the vagina is often underdeveloped and no uterus is formed. Babies with this condition are invariably raised as girls.

HERMAPHRODITISM
Other chromosomal abnormalities have fewer physical effects. A woman with an extra X chromosome, for instance, may never be aware of

it; men with an extra Y chromosome may just be very tall. But sexual disorders broadly termed 'hermaphroditism' do lead to more incongruity between external appearance and genetic make-up. They fall into three categories: female pseudohermaphroditism, male pseudohermaphroditism and true hermaphroditism. True hermaphrodites may have either XX or XY chromosomes, or a mosaic of the two, but develop both testicular and ovarian tissue. The causes of this rarer condition are less clearly understood. Some geneticists argue that rather than two sex classifications, there should be five: males, mermis (male pseudohermaphrodites), herms (true hermaphrodites), fermis (female pseudohermaphrodites) and females. However, there seems little likelihood that such gender identities would ever be accepted.

But we seem to need a categorical statement about a person's sexuality. Anything to do with sexuality and the propagation of the species touches on a very deep nerve.'

Sexuality and gender are so fundamental to our lives that some believe if the distinction between male and female is called into question, society will be thrown into confusion. Such attitudes start at birth when parents ask: 'Boy or girl?' If there is no clear answer, the medical establishment categorises it as a 'psychosocial emergency'.

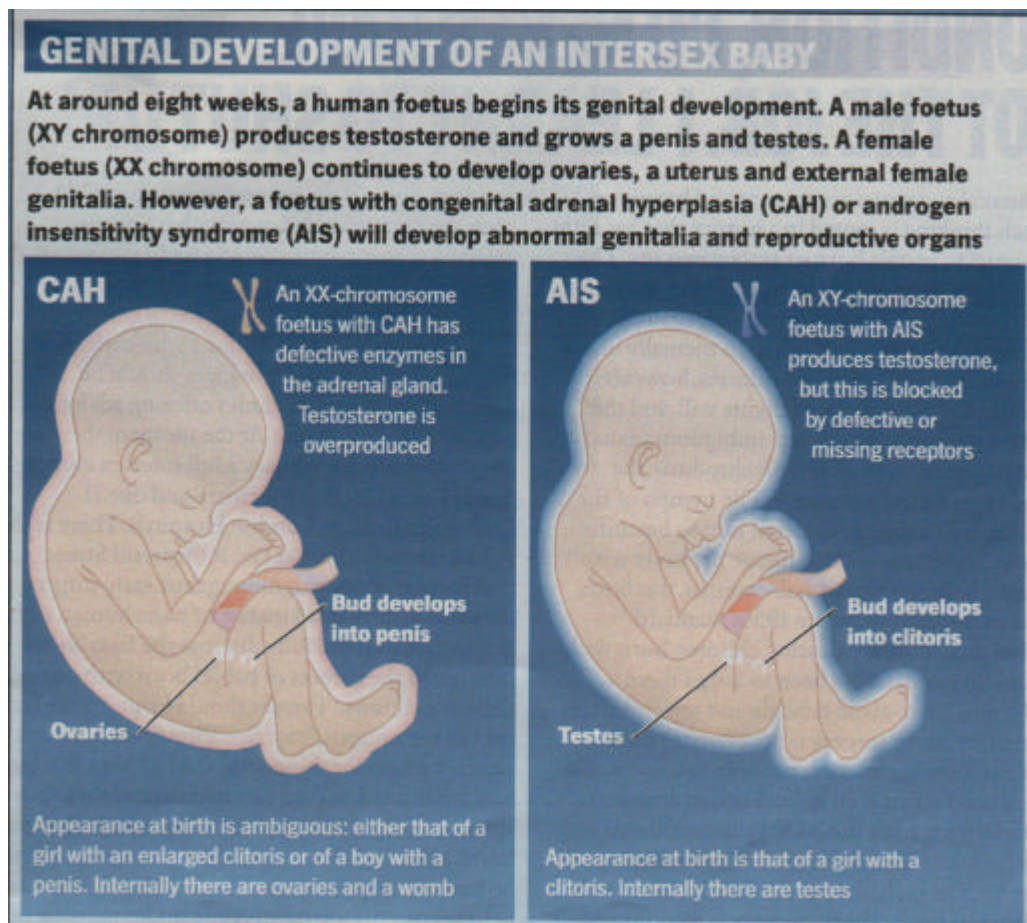
Surgeons, geneticists, endocrinologists and clinical psychologists form a team to decide, usually within a few days, what sex the child is to be designated. The rationale is that parents must be told as quickly as possible to prevent them from rejecting the child. This medical response is based on the assumption that normality is essential for parental and social acceptance and happiness.

Such thinking is rooted in a history that has often been cruel to people born of ambiguous sex. The Romans put them to death to pacify the gods. The Victorians treated some as freaks - relegating them to circus sideshows - and others as mentally ill. In some eastern and Caribbean cultures, however, they are revered as expressions of divine will, and the arts have traditionally cloaked ambiguous sexuality with the Greek myth of Hermaphroditus, the son of Hermes and Aphrodite. The nymph of the fountain of Salmacis considered him so beautiful that she begged the gods to merge her body with his, and a being half-man, half-woman, was born.

In Greek mythology, Hermaphroditus and Salmacis become one body with combined genders



But in the West since the 1950s, standard medical procedure for treating children born with ambiguous genitalia has been to assign them a gender quickly to avoid ridicule and rejection, and perform whatever surgery is necessary to make them conform as closely as possible to a norm. The criteria used is that if an infant's sexual anatomy protrudes away from the body by more than an inch, the baby is considered male; if the protrusion is under 3/8 of an inch, the baby is female. In cases that fall between these criteria, surgery is recommended, and babies are usually made female using the crude logic that, as described in one medical journal, 'it is easier to dig a hole than build a pole'.



The practice of assigning female sex to boys with small penises was pioneered in the United States in the 1960s by a psychology professor, John Money, who argued that nurture was more important than nature and that gender was so malleable in infancy that a male consistently raised as a girl would adapt well to life as a female. He based his theory on the study of a highly publicised case of a boy whose penis was destroyed in a botched circumcision.

The child was given rudimentary female genitals, renamed Brenda and subsequently raised, successfully, Money argued, as a girl. His thesis fell into disrepute when Brenda reached the age of 15 and threatened suicide if doctors refused to reverse the sex change surgery and prescribe him male hormones. He had always felt he was a boy, he told doctors who agreed to perform the surgery.

He is now happily married and a stepfather to three children. Since then, a growing number of doctors both abroad and in this country have begun delaying surgery until a person is old enough to have some say in which gender they are assigned. In some instances, they argue surgery is not necessary.

Support groups in this country for those with intersex conditions believe there should be a network of specialist clinics offering advice on surgery and counselling. At the moment there are only two multidisciplinary adult intersex clinics, one at Leeds General Infirmary and one at University College London Hospitals. There is also a child intersex clinic at Great Ormond Street.

Apart from campaigning against early surgery, they also believe it is a matter of basic human rights that doctors should fully disclose the facts about intersex to the parents of babies born with one of these conditions. 'Parents should be given the facts, told all the options,' says Sue Elford, who chairs a support group for those with CAH. 'Very few are told anything. They are just told their child needs a small operation. It's a one-off. The child will never know anything about it, and everything is going to be hunky-dory; and this is just not true.' Elford believes parents too often come under pressure from doctors to allow surgery to be carried out on their child. 'If parents know they have the option to leave things alone and see how the child matures, a lot more would feel comfortable with this.'

A growing number of doctors share this view. 'We tell children when they are dying of cancer. Intersex is not life-threatening. It is completely illogical not to tell a patient about their diagnosis,' argues Catherine Minto, a clinical research fellow at UCLH. The culture of secrecy that surrounds intersex also means that those not directly affected usually know very little about it. 'People often confuse those with intersex conditions with transsexuals, which they are not at all,' stresses Sarah Creighton.

'Transsexuals are people who feel they should have been a male or a female but have no genetic or anatomical problem at all, whereas intersex people have medical problems caused by a mismatch in their genetic make-up.' Such ignorance, confusion and prejudice means that those who do reveal they are intersex risk humiliation and discrimination; some report being beaten up, others have lost their jobs. 'A common misconception people have when they hear about intersex is to think it means someone has something freakish or deviant about their sexuality,' says Minto. 'The more people know about what it means, the less they will react badly.' Melissa, who is unusual in allowing her real name to be used here, is optimistic that society is slowly becoming more tolerant. She is hopeful of finding a partner and one day starting a family: 'I grew up thinking I'd never be able to have children. Now I know that, with help, I might be fertile.'

The irony is that she sounds masculine. Melissa admits that her gruff voice has sometimes led others to make fun of her, and that her experiences as a child led to many problems as she was growing up. 'I was very shy. I'd rarely go out. I always felt out of place. Because of all the operations I had as a child, I could not bear to be touched for a long time. But I have always liked men. I have had a partner and a sex life,' though she describes that experience as 'atrocious'. Melissa still lives with her parents, but says she would like to get a place of her own, maybe one day go to university. 'I just want a life,' she says. 'I do not feel as if I have had a life yet.'

Anna is more sceptical of society becoming more accepting. On the one occasion she explained her medical condition to an employer, she was met with extreme suspicion. 'I was told that if I had been honest about my condition when I applied for the job, I would never have been taken on. After that, I learnt to shut up.' She asked for her real name not to be printed in this article.

Anna has a feminine voice. She keeps her greying, curly hair cropped short and swept back from her face in a hair band. She never wears a skirt or dress. On the day we meet she is dressed casually in jeans and a sweatshirt. She does not look masculine. She says she does not look on herself as either male or female but, rather, 'mixed gender', and has tended to avoid close relationships. 'Society likes to categorise you, and I do not fit neatly with people's expectations. I just deal with it myself. I rarely talk about it.' She then adds quietly, as if talking to herself: 'It is like being continually punished for a crime I never committed.'

For more information on intersex conditions: www.cah.org.uk (congenital adrenal hyperplasia); www.medhelp.org/www/ais (androgen insensitivity syndrome); www.well-aware.co.uk (general information)