It's a Girl! I Think…
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In Britain there are over 30,000 people living with an intersex condition – meaning they are neither clearly male nor female. Yet this little known group of people have found the medical profession are adding to their ordeal rather than helping them through it, by pressing for unnecessary surgery and by encouraging secrecy and shame.

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Seventeen year-old Samantha recently visited her doctor to find out why she still hadn't started her first period.

She was soon referred to the hospital where an ultrasound scan revealed that she had a few key parts missing: namely a womb, ovaries, and a uterus. That's when she discovered that she was, theoretically, male.

When we ask new parents, "Is it a girl or a boy?" we expect the answer will be straightforward. However, Samantha has Androgen Insensitivity Syndrome (AIS); one of about 75 different intersex conditions which mean an affected person doesn't fall neatly into either the male or the female category. And, so far, the medical profession has been quick to step in and surgically rectify this quirk of nature.

For every 2,000 births, one newborn's genitalia are so ambiguous that immediate sex assignment is difficult. If we also include minor abnormalities - such as undescended testicles or malformed openings of the urethra - under the label 'intersex', the frequency increases to as many as one in every 100 babies, making it more commonplace than Down's Syndrome.

It's only recently that people have begun to publicly question the routine surgery and hormone doses that 'normalise' these babies. Countless research has shown that they grow up feeling confused and violated, and more often than not resent that such an unproven and intimate procedure was done without their consent.

Indeed, the use of surgery to "correct" intersex conditions will be the main topic under fierce debate at the one day Intersex symposium in Turkey next month (May). It also marks the two year anniversary of David Reimer's suicide, the person at the centre of perhaps the most famous sex reassignment case.

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The components which are normally considered to render you male or female are the sex chromosomes (usually XX or XY); the gonads (usually ovaries or testes); and the external genitalia (clitoris, penis etc). A female would normally have the XX chromosome, ovaries and a clitoris, and a male the XY chromosome, testes and a penis.

In intersex cases, one or more of these components is mixed. Samantha, for example, had an XY chromosome and testes, but the external genitalia of a female. She was therefore identified as a female at birth on the basis of her appearance, and has been raised accordingly.

In Britain there are over 30,000 people living with an intersex condition. The most common form is Congenital Adrenal Hyperplasia (CAH), which results in a baby which should be female (XX chromosomes, womb and ovaries), but has no vagina. Instead, an enlarged clitoris gives the impression of a penis.

Melissa Cull, founder of the CAH Support Group, was born with enlarged female genitals, and surgery was chosen as the most appropriate course of action. Throughout her childhood she underwent a number of operations which left her physically and emotionally scarred, and unable to have sex without taking painkillers.

"Even now, corrective surgery is routinely carried out on infants under two years old in the UK," she says, "I strongly believe that any such surgery should be with the fully informed consent of the person involved. Women don't tend to compare genitals (unlike men) so the excuse of embarrassment in changing rooms is a weak one."

For other people, however, their experience of being intersex is marked by the tradition of secrecy and shame that surrounds these conditions.

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Lauren* is unusually tall, with a full head of thick dark hair which bounces off her shoulders. Her skin is devoid of any imperfection, and looks so healthy it glows from across the café room where I am meeting her. It's the sort of unachievable flawlessness flaunted by models in advertisements for foundation powder or Clearasil yet remains elusive for most women. She later reveals that she has never had a blemish in all her 32 years of life, and that this is one of the unexpected side effects of having AIS. The condition is also responsible for her above average height and generous figure, as well as negligible body hair.

Despite her disarming attractiveness, unexpected in someone suffering from an incurable 'defect', Lauren lacks confidence, speaking softly and towards the ground, avoiding eye contact until she acclimatises to the situation. Having been passed around from doctor to doctor, often with little sympathy or explanation, she is curious to find that someone is interested in hearing her story. For her, the struggle all along has been to understand what was 'wrong' with her.

"When I was a child I was just told that I had an operation 'down there'. It was implied that I shouldn't ask questions or even feel the need to discuss it any further. It was as though I was an embarrassing little secret the family didn't want the neighbours to find out about."

Then when puberty became imminent, the story changed: "When I was about 12, and the only thing on every girl's mind was menstruation, I was told that because of The Operation I would never have a period, was unable to have children, and would have to take an assortment of pills and hormones for the rest of my life. That was the entire explanation I got and a 'Well done for being so brave, darling'. Afterwards I just became fixated on what they had done to me down there, it was my little obsession for years."

In fact, Lauren was born with testes instead of ovaries, and it was these that the doctors removed shortly after her birth as they started to herniate downwards. Unlike Samantha's parents, Lauren's were fully aware that she was born genetically male, with XY chromosomes, but it was up to Lauren to find out for herself why she wasn't like the other teenage girls.

In an attempt to overcompensate, she devoured every available morsel of information about menstruation in the hope that her peers wouldn't catch on. She eventually stumbled across her condition when reading about the monthly cycle in a medical dictionary, in a passage that mentioned women who didn't menstruate as anomalies.

"I think the hardest part – other than being lied to and kept in the dark by my own family – was missing out on the whole business of menstruation. It's like a monthly club I could never be a member of or a rite of passage I would never travel through. I would nod and smile when girls whispered about these things to each other, then cry in my room at night while running it over and over in my head. The hormones I was given made my breasts develop but I hated them, I didn't believe that they were real because I knew they were artificially induced.

"I retaliated by distancing myself from my close family, and as other relatives picked up on the tension, my mother was quick to step in and label me a trouble maker. I no longer consider myself a part of that family."

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Ian Fairbrother met similar ignorance in his childhood, an experience which he claims scarred him for life. This time, though, the apathy was from the medical profession, and his mother was as confused and misled as him.

During a bath one night when Ian was just six years old, his mother, Angela, noticed that he had already started to grow pubic hairs. She mentioned this to the school doctor during a routine medical check and Ian was referred to hospital for tests.

He was diagnosed with an overactive adrenal gland, which meant his body was producing testosterone in big enough amounts to trigger an early puberty. Ian says, "All of a sudden I was taller than my peers, as though somebody had put stilts on me. Overnight I became huge: I was six years old but wearing age 10 clothes."

He had to undergo repeated tests, and was given steroids to stunt his growth, leaving him shorter than average today. Each time he attended the hospital, he was stripped to his vest and his genitals examined, a process he recalls as being confusing and humiliating. The doctors were unsympathetic to Ian's discomfort, and he was passed around from specialist to specialist, often having to answer the same questions time and again.

Angela asked if there were any organisations they could contact, or other parents who had experienced the same thing, and was repeatedly told 'no'. This led the family to believe they were on their own with this condition.

In fact, it wasn't until nearly a decade later that she came across a magazine article detailing a condition that seemed very similar to Ian's – CAH. She asked the doctor if this is what Ian had, and the diagnosis was confirmed.

Angela says, "I was confused and angry. For 10 years we had been groping in the dark without any help when all along there was a parent organisation not 20 miles away from our home. The people there were able to answer our questions and most important of all make Ian realise that he wasn't alone with this."

However, the realisation that the doctors had withheld this information added towards Ian's agitation towards the medical profession. He felt that he had been let down and that the focus at the time was on teaching other doctors about this rare condition, when it should have been on providing the best care and support available.

Even now, at 29, he cannot speak of his memories without quickly becoming angered. In particular, he is furious about the photographs that were taken when he was still a young boy.

"I will never forget the photographs," Ian insists angrily. "I was told to strip right down and stand there totally naked. Even a six year old kid knows that it's wrong to be naked, wrong to have your genitals touched, prodded and studied. I resisted the doctors at first, but was repeatedly told I had to do it, had to stand on a box and stretch my arms out either side. An older doctor started pointing the camera and taking photographs over and over again, each time winding the camera forward before focusing on my genitals again. Then I was told to stand sideways with my hands behind my back, and they took several more. I kept glancing at my mum, hoping for support from her which never came, she just stood there silent.

"As soon as he let me go I ran over to her and collapsed in her lap crying, just sobbing for ages. I hated her for letting it happen but at the same time needed her comfort. After that I was desperate to go home, but was forced through more tests, x-rays and examinations. For the rest of that day I barely stopped crying, and wriggled constantly when the doctors tried to take blood or make me stand still for anything."

Ian's mother Angela recalls, "A nurse came and told me Ian was going to have some pictures taken. I assumed they meant x-rays, as I knew he was scheduled for some. I was in another room answering questions about him for a while, then entered the room where they had taken him. To my horror he was standing on a plinth having his photograph taken, naked. When I asked why this was necessary, I was informed it was for teaching purposes. But no permission was requested or given, and to this day I cannot forgive myself for not reacting to this invasion on my son."

To this day the fact that those photographs were taken has been Ian's biggest grievance. When they mentioned these to the support group, they were told they could ask for them to be destroyed. Aged 15, Ian wrote an angry letter to his doctor, demanding that these photographs were handed over in person so that he could see to their destruction himself.

"The doctors knew how traumatised I'd been by those photographs, time after time they're mentioned in my notes. Not once did any of them suggest I get them destroyed, and that this might go some way to compensate for having them taken.

"I was particularly furious with one doctor who had lied about them, even saying none had been taken. He had died some years earlier and now had a ward named after him. Now I can never have a chance to make him answerable for his actions.

"I arranged to pick them up myself, after back and forth correspondance when I was repeatedly told they couldn't be found, among other excuses. When I finally got them I flipped through them and was more shocked than I thought I would be. My face had not been blacked out, as I had been assured it would. I was completely naked, and there were shots of my face and close-ups of my genitals. No 15 year-old should have to go through that. I took them home to show my mum, and we burnt them together."

Angela adds, "At one point, when Ian was still a child, I paused to count how many doctors, students and consultants had seen him over the past few years, and came to a total of 22. Yet even with all this attention nobody bothered telling us the official name of the condition, or offered support of any kind."

American author and Pulitzer Prize winner Natalie Angier writes, "People with [these conditions] do not exist to instruct a benighted world, and some resent being regarded as genetic anomalies that clarify genetic commonness, resent being the ones in the doctor's steel stirrups, being the ones whose faces are blotted out in textbooks but whose bodies are naked and available for public scrutiny."

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In a recent episode of *Footballers' Wives*, character Jackie Pascoe is told her son Paddy has been born with two ovaries and a womb, as well as a penis. The father's immediate reaction is crude: "Let's find a doctor who's got some guts, who'll chop out his girl bits".

The doctor at the hospital gently explains, "Paddy's external organs have developed as male but his internal organs are female. Surgery would mean assigning Paddy a sex he doesn't necessarily have, which could make things much more complicated in the long run".

Cathy Falvey-Brown is an experienced staff nurse and community midwife, who has worked at both Barts and The London NHS Trust and Great Ormond Street. She says, "I looked after a 12 year old asian girl who had been sent to our metabolic clinic as her parents were concerned that she was not behaving normally. They had flown in from India to see one of our specialists, who examined the girl and put her in for a chromosomal test. When the results came back, it was discovered that she was actually intersex, with undescended testes inside her abdomen.

"This was a major dilemma for the family, as the girl would be sterile which carries a huge stigma in their culture and would cause problems should she get married.

"Another youngster came to Great Ormond Street because he was born with a rare condition called exophalmus, in which the contents of the abdomen are on the outside of the body. This is easily treated by surgery, but while he was with us, a paedatrician noticed that the baby had a very small penis and undescended testes. After tests, the baby was diagnosed as being intersexed, and the mother was advised to bring up her child as a girl instead. She struggled with this concept, and found it difficult to consider telling her friends to 'scrap the previous announcement, it's a girl', change his name and have him castrated. In the end she went to the United States to get a second opinion."

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Surgery on the genitals, whether to reduce an enlarged clitoris or to reassign a young boy with a micropenis as a girl, is primarily cosmetic. It has long been believed that 'normal-

looking' genitalia are essential for the development of a stable gender identity. However, this approach has not been supported by research, which instead suggests that surgery is often unsuccessful, both cosmetically and functionally.

In the USA, it has been common practice to operate on any child with ambiguous genitalia since the 1950s, and doctors in the UK quickly caught on. Unfortunately, as consultant gynaecologist Sarah Creighton points out, whether a clitoris is considered large or not is a completely subjective matter. Also, as the body changes during puberty, a larger clitoris is less noticeable, and there is a chance the patient would rather wait and see, instead of risking the loss of sensate function. In addition, follow-up surgery at puberty is often needed, and this only increases the chance of damaging vital nerves.

Indeed, a recent study by researcher Catherine Minto concluded, "Individuals who have had clitoral surgery are more likely to report a complete failure to achieve orgasm.

"Infants and young children are powerless to oppose any procedures, so genital surgery for them is not just a medical issue but also a moral one. On the other hand, some people believe that growing up intersex without a surgically assigned female or male sex can be a very difficult experience for a child."

Researchers at St Peter's Trust used a GenitoSensory Analyzer to assess the sensory abilities of women who had had genital surgery at a young age. It was found that warmth, cold, vibration and light touch sensation was weaker in women who had had surgery than in normal controls. These women also reported a higher incidence of sexual difficulties.

Another study, conducted by May et al in 1996, compared women who had CAH with diabetic women, and found that the former group experienced more pain during intercourse, were less likely to masturbate or attain orgasm, and were still unhappy with their genital appearance despite the surgery.

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Samantha has chosen against having her testes removed, although had her condition been discovered at birth, that choice may have been taken away from her.

This emphasis on immediate surgery has its basis in research done four decades ago on a single set of male twins, Bruce and Brian. Born in Winnipeg in 1965, both boys soon experienced problems urinating, and by eight months a diagnosis of phimosis was given with doctors recommending a circumcision. The inexperienced surgeon and his anaesthetist inexplicably chose a Bovie cautery machine for the procedure instead of the usual clamps and scalpel.

Bruce, who went first, had his penis burnt irreparably, and the damage was so severe his entire organ was destroyed. After this, Brian's circumcision was quickly cancelled, and he recovered without any treatment, making the whole situation even more distressing.

The distraught parents sought the advice of John Money, a psychologist at Johns Hopkins University in Baltimore, Maryland, and a pioneer in the new field of sex change surgery. Harvard-educated Money enthusiastically endorsed the belief that nurture, rather than nature, is central in a child's gender development. He argued that our gender identity is shaped by our upbringing, not our genetic makeup or hormones; and that Bruce would suffer more if left physically defective. Apparently the ridicule he would probably endure in the boys' locker room was reason enough to take immediate action.

Money argued that Bruce's penis could not be replaced whereas a vagina could more easily be constructed. He therefore advised that the boy be surgically reassigned and raised as a girl, and that no-one should ever reveal the truth to the child.

For Money, Bruce offered the ideal experimental conditions for him to test his theories on: a normal, healthy child with a genetically identical twin. In return, he seemed to offer the Reimers their only chance of raising an infant capable of heterosexual intercourse.

And so Bruce became Brenda. At 22 months of age, surgeons removed his testes and reshaped his scrotum as a vulva. Money continued to visit Brenda to assess the outcome, and hailed the operation as a great success in a series of updates and reports he published throughout the 1970s.

Then Brenda slipped out of the public eye until 1997. The reassignment hadn't gone to plan. From a young age, she tore up her dresses, urinated standing up, and was bullied continuously by her peers. After 14 years of discomfort, fighting the gender role that didn't seem to fit, her parents revealed her true sex. Within a matter of months, Brenda decided to become David. He underwent numerous operations, including a double masectomy to remove the breasts hormone therapy had given him, and two phalloplasty operations, followed up with testosterone injections.

At 23, he married a woman with three children from previous relationships and become a stepfather. Throughout their relationship he was plagued by depressive moods, angry outbursts and doubts about his performance as a husband.

A long time rival of John Money, Milton Diamond had always wondered what had happened to the famous twin, especially after Money stopped publishing follow-ups. Diamond, a psychologist at the University of Hawaii, eventually traced David and revealed to him that Money had been using the case to encourage surgeons to continue reassigning infants. Diamond convinced David to go public with his story. After Diamond's report, author John Colapinto covered David's story in depth in a book, *As Nature Made Him*.

Although the book's profits were split 50-50 between the author and David, giving him some financial security, his life was falling apart in all other areas. After being made redundant he never found another permanent job, seperated from his wife, lost huge amounts of money in a bad investment and his twin brother died folllowing an overdose on antidepressants. On 4th May 2004, two years after his brother's death, he used a sawed-off shotgun to end his own life.

It is unfortunate that no-one at the time took any notice of a careful study carried out in the US in 1951, investigating 250 people with intersex conditions who were left untreated as babies. The study concluded that the condition of the genitalia plays an utterly insignificant part in a person's gender identity and self-image. If it had been published or distributed, this research might have averted the generations of genital mutilation that followed. Ironically, the study was done by none other than John Money himself, as part of his PhD dissertation.

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However, Melissa Hines, Director of the Behavioural Neuro-endocrinology Research Unit at City University in London, says, "I think it's wrong to conclude overall that this

practise has been a bad one. The alternatives have been largely untested, and in most cases the children are content with the sex chosen for them.

"I've seen it happen: adults grow up fine and adjusted. It's amazing that it can happen because we think of our gender identity as something really central and not something malleable, something that's always been in us. And what this suggests is there can be a huge social influence on what our gender identity is. So you can take someone who's genetically male – and this has happened where people have had accidents, causing damage to the penis for instance – and they have been reassigned at a young age and lived happily afterwards as a woman."

Last year, Hines was awarded an $810,000 grant from the US National Institutes of Health to fund her next five years of research into prenatal androgen exposure and the development of gender identity and other gender-related behaviours in girls and boys.

However, Melissa Cull is apprehensive: "Much of Hines' research is way out and off the wall, and I don't think she paints a real picture of people with these conditions. I'd like to know why someone in her position is so adamant on early surgery, instead of giving the person with the condition the choice to choose what and how much surgery is undertaken, especially as it can be so risky, both physically and psychologically. Does she really think surgery makes women more acceptable to potential partners? Because it doesn't, with or without it you are outcast and stigmatised.

"Society needs educating that not everyone fits into category A or category B. We need to accept differences and understand that life is precious whatever part of the continuum you are on. Surgical and psychological gender reinforcement wasn't ever the answer to these conditions."

Meanwhile, Samantha remains optimistic about her future. And even though she'll never be able to get pregnant, there is a bizarre possibility – thanks to modern science and the testes she chose to keep – that one day she might actually be able to *father* a child.

What is CAH?

Congenital Adrenal Hyperplasia is an inherited condition which some children are born with (*congenital*). It leads to an enlargement (*hyperplasia*) of the adrenal glands.

The adrenal glands sit atop each kidney, and consist of a inner part, responsible for producing adrenaline, and an outer part which produces three main hormones called steroids. The production of adrenaline is as normal, but the adrenal cortex and its steroid production are at the root of problems caused by CAH.

The cortex produces cortisol, which is responsible for helping the body to deal with stress and for regulating blood suger levels; aldosterone, which regulates salt levels in urine; and androgens, which are male sex hormones. The production of the first two of these hormones is affected because the enzymes required to make them are missing or defective.

The pituary gland in the brain senses the low levels of the hormones and secretes another hormone in an attempt to stimulate the adrenal glands to compensate. However, this constant stimulation ends up causing more androgens to be produced, because these are made normally, and leads to thicker, larger adrenal glands.

Androgens are the hormones responsible for penile growth, facial and body hair, muscle and skeleton development and the deepening of the voice. In males, excessive amounts of androgens cause early sexual development, and in girls they cause masculinised genital development, hirsutism (excess hair), acne and irregular periods. In addition, the lack of salt-regulating aldosterone can lead to dangerously high levels of salt loss in urine, which is potentially fatal.

CAH can be handled with hormone therapy, but there are important ethical issues regarding the use of surgery to correct masculinised genitals in infant females, and there have also been problems caused by a tradition of secrecy and ostracism towards women with CAH.

*For more information about CAH, visit **www.cah.org.uk** or **www.ahn.org.uk**.*

What is AIS?

Androgen Insensitivity Syndrome (AIS) affects people who have XY ('male') sex chromosomes but whose tissues cannot respond, either completely or partially, to male hormones (androgens) such as testosterone. Androgens are the hormones responsible for penile/scrotal growth during gestation, and the development of facial and body hair, muscle and skeleton, and the deepening of the voice at puberty.

There are two forms of AIS, complete (CAIS) and partial (PAIS), which lead to differing body types. In CAIS, the child is born with external genitals that look completely normal for a female, but internal examination reveals that all of the internal female reproductive organs are missing except for the lower vagina. Because of the normal outward appearance, CAIS often goes undetected until puberty, when the person fails to menstruate.

In PAIS, affected children are born with masculinised genitals, ranging from a slightly enlarged clitoris to a near complete penis with scrotum. Some of these children are raised as girls, some as boys, meaning a difficult decision often needs to be made regarding gender of rearing soon after birth.

AIS is usually caused by an abnormal androgen receptor gene. This abnormality renders cells insensitive to maculinising hormones (androgens), meaning that masculine development is impossible in CAIS, or compromised in PAIS.

Normally, although both sexes produce both male and female hormones, those correspondant to the person's sex are made in greater quantities and inhibit the others. In AIS, the oestrogen receptors are perfectly normal and some of the excess androgens get converted into extra oestrogen, leading the CAIS individual to grow breasts and develop a feminine body shape at puberty.

There is some debate about whether the testes in AIS individuals should be removed, as these have a 9% chance of becoming cancerous after puberty. Another issue is whether, in childhood, surgery should be carried out to reduce the size of the clitoris in those with PAIS who identify as female, or to enlarge the vagina.

*For more details about AIS, visit the AIS Support Group UK website at **www.aissg.org**.*