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Ethics and gender

by Lisa Melton

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In the past, children born intersexed were treated with surgery. Now some doctors believe such treatment may be unnecessary.

When Melissa Cull was born it was impossible to tell whether she was a boy or a girl. A small and sick baby, she almost died before doctors discovered what was wrong. After three weeks of tests and X-rays, her parents were told that Melissa was female but had been born intersexed

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due to a rare metabolic disorder called Congenital Adrenal Hyperplasia (CAH).

Cull remained hospitalised and dangerously ill for three months. Though being intersexed is not in itself life-threatening, she had inherited the salt-losing type of CAH that can be fatal if untreated. To survive, she would have to be on steroids for the rest of her life. But as soon as the steroids kicked in and Cull was out of danger, the doctors focus returned to her genitals.

Her clitoris was unusually large, resembling a small penis. The medical team advised her parents that surgery would make her genitals look more female, and the sooner it was done the better. So in 1972, at the age of four, Cull underwent the first of a string of operations designed to "normalise" her.

"It was scary having to stay in hospital, not knowing what was happening," she recalls. "Later, I learnt that this first surgery consisted of the removal of an overgrown clitoris."

The interventions continued. At 12 and 14, Cull had vaginal reconstruction surgery to drain menstrual blood, and enable her to have intercourse later. Throughout the procedures, her condition was never explained. "Each time I went in for surgery, I could get no sense out of the doctors," she says. "It was quite a shock when I found out the details." But the outcome was not as promised. "It was a mess."

As many as 1 per cent of all births involve some sort of malformation, although the majority are mild, such as undescended testes, or hypospadias (where the urethra opens in the wrong place). Cases of sexual ambiguity, where the genitals have elements of both male and female sexual anatomy, are about 1 in 3,000.

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Cull's condition, CAH, is probably the most common intersex disorder. Those afflicted are genetically female, with two X chromosomes. But because their adrenal glands malfunction from early in the womb, they produce masses of masculinising hormones.

Though most CAH women feel decidedly female, and can have children, their early exposure to testosterone leads to an enlarged clitoris and fused labia. "I've always felt feminine, even though my androgens were high as a child," says Cull, who works with computer databases in a Midlands university medical research team. "I can service my own car, and my hobby is model-making, but I also enjoy cooking, and tapestry. The only wiring that went male is my spatial ability."

At 18, Cull received an information pack from a support group that threw her into turmoil: it was the first time her intersex condition had been identified to her. "I grew up thinking that having a family was impossible," she says, "and I put marriage to the back of my mind. It was only years afterwards that I found out that though fertility is much reduced, pregnancy is possible."

But the visible scarring and the pain resulting from repeated surgery have damaged her sexual confidence. "I have always liked men, and I have had a partner and a sex life. But it was atrocious, the guy dumped me because of my condition. It is a difficult thing to explain to anybody with whom you want to get intimate, but I hope, one day, to marry." Now the adult-support co-ordinator of the CAH group, Cull cannot help but wonder what her sex life would have been without that first operation in which her clitoris was removed.

"Because of the surgery, you get total loss of sensation," she explains. "If I'd had the option, I wouldn't have had the clitorrectomy. That is barbaric. I could have lived with an enlarged clitoris. I'm not ashamed to be intersexed, and society should not try to make you feel ashamed."

Today total excision of the clitoris is rare. Surgeons trim the enlarged tissue to an average size, preserving the glans area, which is rich in nerve bundles, for its sexual function. But Laurence Ramecroft, a paediatric surgeon at the Royal Victoria Infirmary in Newcastle, acknowledges that clitoral recession is still a mutilating procedure. "The result of early surgical treatment is not as satisfactory as surgeons would like to think," he says. "There is a movement for doing as little as possible until the child is old enough to be involved." Ramecroft heads a working party that is drawing up a set of guidelines to aid paediatric surgeons and endocrinologists in intersex management.

But is it ethical to allow a child to grow up with ambiguous genitals? Most clinicians argue that such children, deprived of corrective surgery, will be teased by their peers and grow up feeling inadequate and lack self-esteem.

The latest research confirms that these assumptions are not necessarily correct. Sarah Creighton and Dr Catherine Minto, gynaecologists at University College London Hospital, have recently completed one of the few long-term follow-up studies of intersexed women. They compared the outcome of a small group who had escaped surgery with others whose genitalia had been feminised. To Minto's surprise, adult women who retained their intersexed condition did not have overwhelming problems because their genitals looked different. She says: "Some of the best-adjusted women I've seen are in the group that didn't have surgery."

Creighton comments: "Both groups had sexual dysfunction, but those who had surgery had worse problems. The main difference is that of those whose clitoris was operated on, one in four could not reach orgasm."

These findings suggest that it may be wise to delay clitoral surgery, in mild to moderate cases, until the child is a teenager or old enough to decide for

herself. An enlarged clitoris causes no pain to a growing child and surgery is only for cosmetic reasons.

The same 'wait and see' approach could apply to vaginoplasty, a procedure in which surgeons construct a vagina with either a portion of the patient's own gut or with skin flaps. The aim is to make the genitals look as female as possible and to enable sexual intercourse. Surgeons claim that early intervention guarantees the best results and will avoid further surgery. As a result, most intersexed children face major reconstructive surgery by the time they are 12 months old. But the UCL team found, after examining girls between seven and 19 who underwent vaginoplasty as babies, that 77 per cent needed further major surgery. Since constructing a vagina is usually solely for the purpose of sexual intercourse, there is a strong argument for not operating on babies, and waiting until adolescence or beyond.

Though inadequate hormones lie at the root of many intersexed conditions, the causes remain a mystery. Recently, Dr Eric Vilain, of the University of California in Los Angeles, found that an extra dose of a gene named Wnt-4 could be responsible for an XY female with ambiguous genitals.

He told a meeting in London: "This gene is normally present in two copies. If you have an extra copy and you are an XY you will not develop normally as a male but feminised."

Vilain predicts that understanding the molecular mechanisms of how sex genes make a man male and a woman female will be invaluable in assigning newborn babies the right gender and, in future, provide the basis for new treatment.

Dangers of intervention

In one in 50,000 births, boys who are genetically male with a X and Y chromosome) are born with an undeveloped penis, shorter than 2.4cm on average. In some cases they lack a penis altogether. It is technically complicated to rebuild a penis, so the medical approach is often to reconstruct such individuals as females and rear them as girls. Before they are a week old, surgeons create a neo -vagina and castrate the testicles to align the person cosmetically and hormonally with a female. For the past 30 years, it has been widely believed that baby boys reassigned as female early on will know nothing about it, and that parents can rear them successfully as girls. Dr. William Reiner, of the John Hopkins Childrens Centre in Baltimore, vehemently disagrees. "You can do anything you want surgically to a newborn; it isn't going to change who they are, If you have a Y chromosome and you have a normal prenatal environment you are male" he says.

Reiner has followed genetically male babies who, at birth, were surgically reconstructed as females. He found most of the switch back to male gender between five and 16 and show strong male behaviours. What would happen to these children with a micropenis if left untouched?

A common belief is that it is not possible to be a well adjusted male with a small penis. Reiner's follow up studies of children who grew up with a micropenis refute the notion. Not only are they happy being male but some of them are also married. " Males without penises do not kill themselves," he says. "That's a myth."

Intersex links

www.cah.org.uk Congenital Adrenal Hyperplasia Support Group 01543 252961

www.medhelp.org/www/ais/ Androgen Insensitivity Syndrome Support

Group

www.uclh.org/services/repodev/UK UCLH Adult Intersex Clinic

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