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Intersex—seeking the beauty in difference

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At 13 years old, Sean Saifa Wall was admitted to hospital with pain in his groin. He says that he was given very little information about what might be causing it, and doctors didn't discuss different options for treatment with him. He was told that his testes had to be removed immediately.

"I remember before surgery… I asked the nurse what was going on, and [she] was saying that I have these gonads that need to be removed. I'm 13 – I don't know what gonads are." The nurse told Saifa that it was because "they're not good". To Saifa it sounded logical: "If it's not good and it's in my body, it probably should be taken out."

But today he still doesn't know what, if anything, was dangerous about keeping his testes or what was causing the pain.

Not too long after the surgery, he remembers one conversation in particular: "The surgeon was talking about how he wanted to create a vagina. The way he described it… it sounded barbaric." Saifa says, recalling that he was sat in the surgeon's room in horror.

"My mum was to my right… and I was probably turning green, and [she] looked at me and said, 'Do you want to go through with this?'" Saifa immediately said no. "I remember the surgeon was saying, 'We're going to shave down the clitoris.' And I was like, this all sounds painful and horrible. I think, in that second, that one moment, that was what spared me from genital surgery.

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As a child growing up between New York and North Carolina in the late 1970s and early 1980s, Saifa was at the cusp of a change in thinking about the medical management of intersex conditions in the USA. He is one of seven living relatives with the same intersex condition. Three of these relatives identify as female, having undergone surgeries in childhood to remove their testes. "That was the protocol of the time," Saifa says.

As it became an option to leave the testes in the body, Saifa's mother decided that he should not have them removed before puberty. "I guess sometimes the more information people have, they're inclined to make different decisions," Saifa says.

He has androgen-insensitivity syndrome (AIS), thought to affect between one and five in every 100,000 people. He has XY (typically male) chromosomes, but the receptors in his body that respond to male sex hormones don't work fully. Saifa's body developed with some male characteristics, such as testes in his abdomen. He also developed some female characteristics, including breasts, when he was eight years old.

Even though there are a number of people with AIS in Saifa's family, he says that they are not able to talk about their shared experiences together: the shame and stigma even reaches into family circles. "You would think that there would be support [in the family], that we would be able to talk about it. But I think it shows that a lot of the thinking at the time prior to my generation was that this was something not to be shared. So there was this vow of silence with anyone else outside of you and your doctor."

Saifa is the only member of his family who openly talks about his condition. Now he works as part of a global network of intersex activists to promote a healthy image of the intersex body, campaigning against surgeries that aren't medically necessary.

"Intersex bodies are beautiful," he says. "People should have the right to choose, to bodily autonomy, to reproductive integrity. But the immediate work to be done is to educate people on what it means to be intersex. That's where we're starting."

Saifa chooses to use the term intersex, rather than differences in sex development (DSDs), to 'own' the traits he was born with. Others prefer to say 'DSD' rather than intersex. Estimates of the number of people affected vary. The Intersex Society of North America says between 1 in 1,500 and 1 in 2,000 babies are born with such atypical genitalia that a specialist is called in, but adds that many more people are born with less obvious variations that might not appear until later life.

DSDs encompass a wide range of conditions that can involve a person's genes, combinations of their chromosomes, hormones, genitals and reproductive organs, and secondary sex characteristics such as body hair distribution, muscle mass and breast tissue. DSD was previously known as hermaphroditism, a term now considered outmoded and offensive.

Some DSDs are extremely rare, others much less so. Some are life-threatening and others cause no medical problems. Some conditions give a person sex traits that are typically male as well as some that are female. Sometimes these traits can't be easily classified and might appear ambiguous or 'in between'. Some people with a DSD may have no ambiguity but just a slight difference in appearance or bodily function.

For many people born with a DSD, the binary system of male and female just doesn't cover it.

The second half of the 20th century is littered with stories of children having surgery done to them just so their bodies conform with expectations of what female and male genitals ought to look like.

Surgeries done without full consent – either because someone is too young to understand or because doctors don't disclose enough information – are sometimes acknowledged decades later in the painful process of dragging a case through the courts. Last year in Germany, Michaela Raab sued doctors for removing what they called an "oversized clitoris" and initiating female hormone therapy without mentioning to her that she had XY chromosomes. Raab said this information would have affected her decision of whether to go ahead with surgery and hormone therapies, and the court found in her favour.

But things are changing, slowly.

"It all started in the 1990s when intersex activists, who were former patients, started to come out to say, 'What's been done to us is really not OK. I feel ashamed and I shouldn't feel ashamed,'" says Nina Callens, a clinical psychologist specialising in DSD at Ghent University.

The medical community responded by publishing a statement in 2006, known as the Chicago consensus. Some medical practitioners used this to set a standard of care for people with these conditions. Callens says that good practice includes avoiding medically unnecessary surgery, working in multidisciplinary teams, and providing psychological support for and encouraging more discussions about these conditions with patients and families.

If a child is thought to have a DSD, the first step is that the family is referred to a centre with expertise in that area. "What typically happens is they run tests, but the test results don't usually come back in a few weeks," Callens says. The period of limbo can last for a long time. The sad part, she says, is that for more than half the children with a DSD who have XY chromosomes, a genetic cause – often required for a firm diagnosis – will never be found.

A lack of knowledge around DSDs as well as the associated taboo and secrecy makes talking openly about the conditions a hugely important part of treatment. "There are a number of concerns that we have to help [families and people with DSDs] deal with. That takes an awful lot of listening," Callens says.

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The Chicago consensus, while perhaps a list of good intentions, does not seem to have percolated down to frontline care in many cases. The lack of psychological support for parents of children with DSDs is particularly stark when considered alongside the recommendations outlined in the 10-year-old consensus document.

"I think the medical side of things is phenomenal, but the emotional, psychological side of things is rubbish," says Natalie (names have been changed), a mother of a child with a DSD in England. Even at the larger, specialist hospital Natalie went to, she felt that she did not receive anywhere near the level of support needed when she and her husband had a baby with a DSD.

"It was a textbook pregnancy to start with," Natalie explains. Seven months into their third pregnancy, she and her husband decided to find out the sex of the baby. "We went for the scan, very excited, and I could see – having had lots of previous scans – that the sonographer could see the baby's genital area. What I couldn't work out was what she was looking at."

After several consultations, a geneticist explained to Natalie all the reasons that they were aware of for the ambiguity that the sonographer had found and some of the possible conditions the baby could have. "We were left a bit bewildered about that because obviously they were all of the things that could happen to our baby, not what would actually happen to our baby."

The ability of doctors to diagnose DSDs and perform genetic testing is advancing rapidly, but the emotional management of the conditions hasn't received the same focus or investment. Natalie faced avoidable problems. Despite all the information on her child's condition in her medical notes, which she reminded various health professionals to read, they struggled to find ways to talk about her baby. Even the hospital's birth database wouldn't accept an entry unless the child was assigned female or male – which hadn't yet been done for Natalie's baby.

With an overload of scientific information but a lack of helpful psychological support, Natalie's main concern was whether she would bond with her baby. "Sex and gender isn't important until there isn't one," she says. But when her child was born "blue and floppy", Natalie knew straight away that it didn't matter at all. "I just wanted him to breathe – and he breathed, and it was fine."

Natalie's son was born with XY chromosomes, an underdeveloped uterus, a half-formed 'streak' ovary (with a fallopian tube) on one side and an ovotestis (made up of both ovarian and testicular tissue) on the other. He also had hypospadias, where the opening that is usually at the tip of the penis is found on the underside. It's a common DSD affecting between 1 in 200 and 1 in 300 boys.

At eight days old, Oscar was assigned male. Natalie was told that his reproductive tissues would never be capable of producing an egg or carrying a fetus, and at nine months they were removed as they were said to pose a high risk of causing cancer.

Oscar, now two years old, is partway down a long road of surgeries. The operations he's had so far haven't all been straightforward. He suffered sepsis after one and also has bladder problems unconnected to his DSD, which will mean further surgery in the future. But the operations Oscar has had, and will have in the near future, are all medically necessary, Natalie says. She's not interested in cosmetic procedures to make him appear more typically masculine.

Debbie and Jack's child was just a few hours old when a doctor walked around the curtain of the bay and asked for the unofficial birth certificate back. Debbie had been given the token soon after giving birth. "She said, 'We don't know if the baby is a boy or a girl,'" Debbie remembers. "It was pretty blunt, to say the least."

Like Natalie, Debbie found the way medical professionals spoke to her about her child – let alone the lack of active support – very hard to deal with. "Constantly in the early days we were met with the terms 'abnormality' and 'birth defect', and it's very hard to have someone repeatedly telling you that there's something wrong with your child, that they're somehow not normal, not perfect."

Over the next few days, doctors examined Freddie for various DSDs, including congenital adrenal hyperplasia (CAH), a genetic condition that affects boys and girls in equal numbers.

CAH is a condition in which the body can't produce certain steroid hormones, such as cortisol, which can cause a build up of androgens – sex hormones that give rise to typically male physical characteristics. Children with CAH who have XX (typically female) sex chromosomes can develop ambiguous genitalia, such as an enlarged clitoris that resembles a penis. Children affected can also have abnormal salt levels in the blood, which can be life-threatening.

Debbie and Jack left the hospital when Freddie was four days old. Although they had the birth certificate back, they were advised not to register Freddie's birth until they had some more concrete answers.

Initially a team of specialists at a children's hospital seemed much more able to talk to Debbie and Jack openly. After a very brief look at Freddie the surgeon said that nothing needed to be done immediately as the child wasn't at any immediate risk. Then the endocrinologist shared some initial test results. Freddie had XY – typically male – chromosomes, but they needed to wait for the full tests to come back to know more.

"It was very difficult obviously, at that point, for him to tell us anything. He just told us what we needed to do for various tests," Debbie says. "We didn't know whether he could produce testosterone, we didn't know whether he would react to testosterone. There was a lot of uncertainty."

The tentative results edged Debbie and Jack closer to the information they wanted, but they still had to live with big gaps in their knowledge. They still don't have a firm diagnosis. Debbie asked whether any counselling or support groups were available, with no luck. "Nothing was ever offered, and when I asked, they couldn't point me in the right direction."

Medical professionals have a responsibility to talk to patients about both sides of the story, Callens says, to provide all the medical information available while being open about things that just aren't yet known or understood. This is especially important in the long term. Having – or not having – this kind of information can shape how a parent forms a relationship with their child. It may play a part in making decisions about how to bring up the child. "I think the medical community sometimes does a little too little listening," she says.

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Last year, Malta passed legislation that made it illegal to perform 'normalising' surgeries on children with DSDs. The law was greeted by campaigning organisations across the world as a landmark in the recognition of rights of people with DSDs, and a huge step forward for Malta.

This came as part of a bill to ensure that everyone – including transgender people and people with DSDs – is adequately protected by law. People with intersex characteristics might not be at either extreme of the male–female spectrum biologically, but the idea that they are 'trans' – where one's identity and bodily sex characteristics do not fully correspond – does not necessarily follow.

Most of the medically unnecessary interventions happened for social reasons, rather than from immediate medical need, says Silvan Agius, who has worked in intersex and LGBT rights for many years, and is now director of human rights and integration at Malta's Ministry for Social Dialogue, Consumer Affairs and Civil Liberties. "The doctors themselves often speak about the social dimension: how will this child grow in the world where there are males and females and they will be unable to go to the public toilet and pee like all the other men, for example. Is that a valid reason to intervene on a child's body?"

Earlier this year, Ed Clere, a Republican member of the Indiana House of Representatives, proposed a similar – if narrower – bill at the US state government level. Clere was struck by a court case in South Carolina, in which the adopted parents of a child with a DSD are suing the hospitals where sex assignment surgery was performed on their child while he was in foster care. The child, known as MC, identifies as a boy, but during the surgery his penis and an undescended testicle were removed. MC's adopted parents argue that the surgery was not medically necessary and will have a huge impact on MC's ability to reproduce and express himself sexually when he grows up.

In his bill, Clere focused on children in state custody, so as to avoid opening up broader debates over tensions between parental rights and children's rights, although he thinks that will be an inevitable discussion at some point. "What I was trying to do was distinguish between medically necessary surgery and surgery that's performed to make a child's appearance conform to a social or cultural norm," Clere says.

The bill, put forward in January 2016, didn't receive a hearing. "I don't think this issue of intersex conditions is something that's familiar to most legislators," he says. "It's dead for this session." Clere has continued to talk about intersex human rights in Indiana to try to bring the issue up the political agenda. "This is an issue that hasn't received much attention at the state level, although I expect we'll start to see more," he says. "The issue of intersex conditions has a medical and a scientific dimension to it, but it also has a political dimension."

In the UK there is also interest in proposing legislation to protect children with DSDs from medically unnecessary surgeries. The precedent that Malta set is very encouraging, says Lord Stevenson of Balmacara, a Labour frontbench peer born with hypospadias and a founder and trustee of the charity Hypospadias UK. Malta's law is "a ray of light in an otherwise pretty bleak world," he says.

"Now that we've got a better understanding of what's happening in the small but very important number of people that this affects, then we need accommodation there. I think there's room to amend existing legislation to try to do something about that."

The clause passed in Malta to prevent unnecessary surgeries on children with DSDs was incorporated into a bill focusing on transgender rights. What do these kinds of links to transgenderism mean for those living with DSDs?

Transgenderism and intersex are very different, Silvan Agius says, but they are related. "The basis for the discrimination is the same," he says. "The gender binary notion of society – that creates the difficulties."

Artificially simple binary systems of male–female simply can't account for the ordinary, natural variation of human bodies, let alone the complexities of human gender identity. "If in society we only had two races you could identify with – let's say people could only identify as white or black – that would eliminate the visibility of people who identify in other ways. This is the same thing that's happening here," Agius says.

Transgender and intersex people face different prejudices. "Interestingly enough, the body is not accepted in the case of intersex people," he says. Here the underlying social assumption is that "nature has made a mistake with them and science knows better."

Agius says that although intersex and transgender people face different challenges and have different needs, in the case of Malta's law it was very much a pragmatic solution to include intersex and transgender rights in the same legislation.

"It's a difficult issue for a lot of legislators," says Ed Clere. It's a balancing act between being heard as part of a louder voice, and being understood as a clear and distinct one. "Given the scant attention that intersex issues receive, I think we should take advantage of any opportunity to raise those issues and discuss them," Clere says. "The LGBT discussion provides that sort of opportunity."

Sean Saifa Wall says that medical practitioners' growing awareness of transgender people and their needs has opened their minds to listen a little more to intersex patients. "Increasingly I come across [medical] providers who respect my decisions as far as wanting hormones and certain surgeries – and I've only had chest reconstruction surgery. That's increasing," Saifa says.

Improved awareness of transgender-specific care among medical providers, however, is a mixed blessing for intersex people, Saifa says. "Transgender-specific care and medicine reinforces the idea that gender identity is confirmed through surgery," Saifa says, and quotes a saying he's fond of: "Intersex people have to fight not to have surgeries that they don't want, and trans people have to fight for surgeries that they do want." That dichotomy, he says, is really devastating.

A lack of specific protocols and creativity around working with intersex patients is a real problem, Saifa says. Theories, practices and protocols that aren't made by intersex people for intersex people means that there is a risk of being sidelined.

"I think there's a lot of work to do," Saifa says. "The protocol has been surgeries and subsequently hormone replacement therapy that reinforces the gender, along with socialisation. I feel like there's not a lot of thinking outside the box with regard to treating intersex patients."

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Human bodies don't all fit the binary system of male and female: the fact is straightforward, but breaking the silence, dismantling the taboo and casting off the shame is not. The silence can seem overwhelming, from maternity wards where awkwardness gets in the way of basic care, to legislatures where protection of human rights slips down the agenda because the idea is simply too unfamiliar.

At the moment, legislation and medical standards fit to care for and protect people with DSDs are a long way off in most parts of the world. But the pieces to make the shift happen – the activists, families, wellinformed doctors, lawyers and legislators – are beginning to come together. It's just as well, as breaking through the stigma will take a lot of momentum.

But the discussion is being pushed forward in wards, ministries and living rooms. Both Natalie and Debbie are trying to keep the conversation going in their immediate family, and are finding ways to try to make sure that their children understand their bodies as they grow up. "I want [Oscar] to be comfortable and confident in his body, and regardless of what's going on," Natalie says. "That is my primary concern."

Differences in general with regard to sex and gender are becoming gradually more widely understood and debated. The more open and tolerant social attitudes to sex and gender differences become, the better Freddie and Oscar's chances of finding acceptance. The hope is they won't face the silence and shame that Saifa and others had to. "It's not freaky, it's just something that happens. It's nature," Natalie says.

"It's not weird – Oscar is a two-year-old who is a two-year-old. He's not a freak, he's not weird, he's not odd, he's not strange, he's just two – he's a child. But it's not spoken about, so people don't get that.

"Children are born like this. That's what people need to understand."

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