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## Working together in placing the long term interests of the child at the heart of the DSD evaluation<sup>☆</sup>

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**Abstract** This paper articulates a number of important but often ignored questions that families have during and following the diagnosis of a child with a DSD. Recounting a personal birth experience, it illustrates the urgent need for more psychological and educational support during and following diagnosis and gender assignment. Finally, the paper describes some practical strategies for raising a child living with genital difference. It urges everyone involved in the care of children with a DSD to support parents in looking beyond initial anxiety and worry and always to place the child's long term wellbeing at the very heart of the clinical decision-making and care. © 2012 Published by Elsevier Ltd on behalf of Journal of Pediatric Urology Company.

"...Finally, there must always be room for the atypically sexed child to imagine and to *become*, of their own accord, rather than to simply respond to what has already been imagined for them." (Katrina Roen<sup>1</sup>)

I am the mum of three great kids, all with their own little character traits that can make being a mum both rewarding and exhausting. My eldest daughter has just turned 10 and was diagnosed at birth with Partial Androgen Insensitivity Syndrome (PAIS). I am also the administrator of a support and information resource for families of children and for teens with a disorder/difference of sex development (DSD), called [dsdfamilies.org](http://dsdfamilies.org).

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URL: <http://www.dsdfamilies.org>.

<sup>1</sup> Roen, Katrina, 2009, Clinical Intervention and Embodied Subjectivity: Atypically Sexed Children and their Parents, in 'Critical Intersex', Morgan Holmes (ed.), Ashgate Press 2009.

In an email sent by the organizer of this conference, he stated his aspiration that each participant should list the most important questions.

I am afraid we parents have rather a lot of questions:

- How do we cope with having to assign a gender of rearing for our own child?
- How do we tell well-wishers that we don't know whether our baby is a boy or a girl?
- How do we deal with having to make fundamental and sometimes irreversible decisions about the care of our children if the doctors who inform us – often the only ones we discuss care and interventions with – are also those to whom we have to give informed consent in acting on those decisions?
- What happens to parents and their affected child following the initial 'evaluation', when we leave the hospital? Or when we leave the DSD clinic?
- Which expert provides continuity of care during those long periods when we do not see the multi-disciplinary

team, but during which time the questions on what to do, what to say, how to deal with this or that... keep coming?

- How do we ask the school to allow our young son to use private changing facilities and toilets, so he will not run the risk of being ridiculed and bullied? Or should we instead, teach our son and those around him about differences in people's bodies, and not seek special privileges for him?
- How and when do we tell our daughter that she cannot grow babies in her tummy?
- How do we prepare our child for a genital examination?
- How do we share information about our child's condition, and instil a sense of privacy without attaching a sense of 'wrong' or 'shame'?
- How does one explain scar-tissue?
- With whom can an adolescent girl talk about sex when her body is different from those of her teenage friends?
- How do we raise confident and happy kids with a DSD?
- How do we place self-esteem, a positive body-image and gender wellbeing at the heart of their care?
- How do we help our daughter understand that having good sex is not the same as having a typical sex anatomy?

My questions, my dilemmas as a parent, are probably very different from yours, as medical professionals.

When focussing on the topic of the DSD evaluation...

Shouldn't the key questions include how to place the long term interest of the child at the heart of that evaluation, and who decides what that long term interest is anyway?

Surely any update on *evaluation* has to be holistic, and include all dimensions of care, not focus in on surgery above all else. Some concepts that I believe need to be addressed include:

- How we describe genitalia (avoiding outdated and insensitive language);
- How we discuss long term outcomes of early surgery with families (both for girls and for boys);
- How we support the family through, and following, a gender assignment process;
- How we offer families a vision of 'normality through acceptance' rather than via early surgery ...

All these things affect the outcome of any evaluation, and indeed, impact centrally on the decision-making process as to whether surgery is the right choice for a family (at any given point in time).

Surgery may be, and often is, a choice that parents make, but whether they are adequately supported to make that choice with the right information – and in the right frame of mind – is still, too often questionable.

I do not have the answers, but I would like to work with you in helping families feel as supported and as informed as possible.

I can tell you about two things reflecting my personal experiences as a parent of a child with a DSD:

The first one is my experience of the process of gender assignment for my child with a DSD. The second thing I can tell you about is raising a child with genital difference.

## Gender assignment

Let me begin by recapping the events following my first child's birth:

My daughter was born following an emergency caesarean section; everyone in the delivery room congratulated us on our beautiful daughter. Some 12 h later, after telling all our friends and family our good news, a junior doctor told me – without my husband being there – that her swollen labia might have testes in them.

A few phone calls later, my mother confirmed to me that at least two members of my family had been diagnosed with what was then called 'testicular feminisation'.

My family had knowingly taken the chance of not telling me about the hereditary pattern of the condition, therefore taking away from me the opportunity, *and my right*, to fully prepare and inform myself about the condition.

Despite giving birth in a large city teaching hospital, it took another five long days before we were introduced to the clinical experts: a paediatric endocrinologist and a surgeon.

During these first five days in the maternity hospital one consultant would contradict the other, leaving us in total despair as to what was going on with our child.

A day after finding testes in our baby, and having identified two 'similar cases' in my family, a consultant radiologist examined our child. He could not find testes and he declared, without consulting his colleagues, that our daughter did not have the condition. 'What about the family connection?' we asked. 'Just a coincidence', was his reply. This was what we wanted to hear, and we did not question his medical authority. A day later, we were informed by another consultant about the karyotype, and we came to understand how credulous we had been. We never received an apology.

During this time we avoided friends and phone calls, and internet research did nothing to give us hope about the future our daughter would have, or to help us understand the situation we were in and what we were to expect.

On day six, at our first meeting, the surgeon comfortingly put his hand on my husband's shoulder. That small act of human kindness still stands out.

I did not want this doctor to go away, but I did not understand why he was there: there was nothing wrong with my child, she was perfectly healthy. She had XY chromosomes, but I knew for sure you could not cut the Y chromosome away – what did a surgeon want to do with her?

I still wish he had introduced himself as 'a urologist' – perhaps then I would have understood his perspective.

The other doctor was 'an endocrinologist'. He too was very friendly, very soft-spoken and empathetic; but I did not have a clue what he did, what he measured, what he looked at – how he was involved in the care of my child.

I was exhausted and bewildered. The doctors had lost me at the first hurdle: the introductions. I did not understand their expertise, nor how they were going to 'evaluate' my child, and so I could not begin to understand what was the matter with my child.

We wanted someone to tell us how our daughter was going to cope *psychologically* with having XY chromosomes. How we were going to cope psychologically with this. There

and then we needed someone to give us some perspective on the medium and long term mental wellbeing of our child.

And I needed someone to *empower* me, to tell me that – even if I didn't understand much of this yet – my/our parenting would play a really important part in my child's wellbeing, and that they would help me and give me strategies to be the best possible mum.

Then the doctors told us that first our baby had to go through a gender assignment process, and that we should delay the registration of the birth. My husband understood, and he cried. I just felt numb.

A few months ago I was asked by a clinician – a urologist – what happened at my daughter's birth. I think this was the first time I spoke about this out loud.

And so I could not stop crying as I was trying to explain how I experienced the gender assignment process: that it feels that the baby you had might have to pass away, and that instead there might be this new baby, with a different sex, whom you'd learn to love too, but still, the child you had would be gone forever.

And the answer from this doctor: 'We don't need five urologists on our team, we need five psychologists'.

Some twelve days after her birth, we agreed with the recommendation to raise our baby as a girl. We were given a final piece of advice: 'don't lie to her, never lie'; and we never have.

As an aside, I was quite shaken when about a year ago a senior endocrinologist told me how shocked he had been 'to read recent research about parental distress at the time of gender assignment, and how indeed some parents suffered from Post-Traumatic Stress Disorder years afterwards.' How I wish I had the presence of mind to say 'How can you not have known this?'

A gender assignment process is for many parents traumatic; but how we – medical professionals and support resources together – help parents at that time can make fundamental differences:

1. to the decisions that are made during and following the DSD evaluation, and
2. to the ability of parents to adapt to the diagnosis and move forward.

I was recently introduced to a mum of three girls, the older two of whom have congenital adrenal hyperplasia (CAH). I told this mum about writing this paper and asked her if there was anything I could raise on her behalf. A few days later, she sent me the following:

I want my 3 daughters to grow up to be happy and confident with who they are. This is not dependent on them having the 'perfect' bodies, if that were the case we would all be miserable! For my two girls who have enlargement of the genitalia as a result of a medical condition (CAH) I believe that there are two key factors in helping them to come to terms with their condition. One is that their parents accept and are comfortable with the physical effects of their condition and the other is that the emphasis of their condition is not placed on these obvious physical effects.

Surgery in childhood to cosmetically reduce the obvious physical effects of differences in genitalia may help

parents to feel that their child is more 'normalised' and therefore help alleviate some of the anxiety and stress. However, this alone does not help parents to come to terms with, deal with the challenges and fully accept a complex and private condition and to know how best to support their child. If parents continue to struggle to accept the condition the child will be very aware of this and will be negatively affected.

It is only at 9 1/2 years old, in the run-up to swimming classes at school, that I have made my eldest daughter aware that her medical condition has affected the appearance of her genitalia, despite having a younger sister who is not affected. In fact it took 4 years before either of my girls with CAH noticed that their genitalia were different than their younger sister's. With the wonderful wisdom of a child my eldest daughter responded to her sister's query about this that: "We are all different!"

## Raising a child with genital difference

This brings us to the second part of my paper: how to help parents imagine and understand that deferring surgery is a realistic option?

New parents are afraid, instinctively want to 'protect' their child and can all too easily equate that to 'normalising' surgery.

But it is my sense that this instinctive response has often to do with not knowing how to deal with practical issues: how to deal with babysitters, and how to deal with nurseries, what happens if someone offers to change your baby, or when you are on the beach? And how to cope with those swimming classes...?

With all the knowledge and evidence we have, surgery should no longer be a treatment to pre-empt bullying, and certainly not a treatment to alleviate parental anxiety, without at least trying first to build better support and management strategies.

I want to share some of these strategies, as it is to you, as medical professionals, that new parents will look for advice and instruction.

## Terminology and language

You may have noticed that I avoid the terms 'genital ambiguity' and 'ambiguous genitalia', which I think are very unhelpful, and suggest 'wrongness', 'something in-between', which really only plays into the 'exceptionalism' that surrounds these conditions and helps to produce secrecy and isolation.

I use, and would recommend the use of, the term 'genital difference': words that are simple and descriptive, rather than pejorative and confusing.

Some families seem uncomfortable to talk about genitals. We have made this a straightforward topic in our family by using child-friendly words. I borrowed an innocent little word for female genitals from a francophone friend – we call it a 'mimi'. My girls have mimis and my son has a willy. When you can talk about mimis and willies openly, it is not threatening. 'Your mimi is a bit different' – that is not scary, not to a child, and not to the parent who has to tell it.

Mimis and willies are private – that is a message that my daughter has heard from a very young age onwards, and I continue to use that very same message now also to her brother and sister.

### **Changing, beach**

I recently read an unpublished interview in which parents argued, against their doctors' recommendations, in favour of surgery because according to the parents it 'made changing your child on the beach so much easier'.

As a first-time parent, you probably have only seen the baby clothes and the pram (baby carriage) sections of the "Babies R Us" store ... You probably don't know that there are wonderful Dora the Explorer or Dinosaur hoodie towels that all little kids love to have on the beach – it is *those* towels that make changing on the beach so much easier.

### **Changing, babysitters**

My advice to parents who consider surgery in case a babysitter might notice the child's genital difference would be something like this:

Take a few months to recover from childbirth, get your feet back on the ground, and take some time to adapt to the diagnosis. Only then are you in a position to consider properly all the options for your child's care.

During that time you learn so much about the diagnosis; it becomes less overpowering and less all-consuming and less frightening. You will have experienced a few awkward situations, like a friend wanting to change your baby's nappy (diaper)... Again, there are many strategies: you can say 'Oh, I really want to change her nappy myself – it is special bonding time for us' or a firmer 'You don't want to do that – each time we remove the nappy she does something; I'll quickly go and do it myself, but thanks.'

Or, take a leaf from my friend Maggie's book and tackle it head-on. She says: 'I had good experiences with just mentioning it at the end of the typical "this is what you need to know" conversation with babysitters (and kindergarten!). I would say: "Oh, and if you are helping him in the bathroom (or putting a nappy on for bed), you may notice that his genitals look a little different. Don't be worried, this is how he was born, and he is healthy and fine." I have had no questions or strangeness from these caregivers.'

### **Nurseries**

The mum of a baby with complete androgen insensitivity syndrome (CAIS) recently shared with me that her 10 month old daughter's labia are very swollen, and despite the reassurances of her medical team she is worried that people at the nursery her daughter will be starting at soon will notice.

Her email reminded me of my own concerns about how other people would view my daughter's genitals. I wrote back and shared Maggie's approach: 'If someone were to say that her labia look a bit swollen, just say that she was born that way, she has been checked out and she is perfectly healthy. Full stop. I do understand how you feel

though – I remember looking over the shoulders of other mums in the baby changing rooms of my local John Lewis department store trying to see how their girls look, and whether mine was very different.'

Next time I write I'll tell her how after changing to a different nursery I spent my daughter's first day at her new nursery PETRIFIED. By that time (age 2.5) my daughter's clitoris had become more pronounced, and with a urogenital sinus one could clearly see 'difference'. When I picked her up I panicked and blurted out to the young member of staff who handed my daughter to me 'she looks a bit different doesn't she'. The young woman did not have clue what I was talking about. The majority of staff at nurseries are young people who have very little experience in 'life' – I felt so silly for having been scared of this young woman, who has totally different things on her mind than what my girl's genitals look like.

The mum of a little girl with ovotestes told me about the support she *receives* from the nursery: 'The director of my daughter's nursery could not have been more supportive, saying 'don't worry – all children are different', and they have always helped me and my husband to treat the issue calmly and with 'normality' – but she adds: 'It is not always easy, living in a small village'.

### **Working together**

Caring for a child with genital difference means you are always on alert, but once you get used to it, and once your child gets older, it is not so hard.

What is difficult is having to do it amid the isolation that can come all too easily from our children's diagnosis, and this is where we parents need your help, as medical professionals, in stimulating contact between affected families, in helping us share these experiences, with each other, with you and with the many parents who are too afraid to reach out to each other.

What is also difficult is that sometimes we don't really know how to care for genitals that are different. When my daughter was around 4, and we were in-between medical teams living in the UK, I was worried about some white discharge. I did not want to ask our family GP (MD). I ended up emailing a physician I did not know personally but whose comments I had read, and hoped he would help. And he did; he replied very quickly, and my family will always remain grateful for that. And as he wrote, the discharge is nothing abnormal, just needs a regular wash.

But when you have no-one to ask, it can turn 'nothing abnormal' into something insurmountable, something scary and something that causes anxiety.

Are there tricky situations? Yes.

When in early January, after the first day back at school, my daughter (then nearly 10) told me 'this term we are having swimming mummy, it's tomorrow', my heart sank, and I thought 'Oh no, not again, I thought we did the swimming in year 4'...

The rest of the afternoon I was really concerned about what I should do: I don't want to make her difference a bigger thing than it is, but equally, I need to 'protect' her, or as I have come to see it 'empower her to protect herself'.

So that evening in the car on the way to the Brownies (Scouts), I said to her 'So, you are having swimming tomorrow?'

'Yeah', she says.

'Do you remember what I told you about keeping private bits private?'

'Yeah mum', and she rolls her eyes.

'OK sweetheart', I said, 'sometimes I don't know if you remember the stuff I tell you, but if you do remember that is fine.'

A minute later, I dropped her off, and that was it.

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