

# **A Call to Action: The Development of a Decision-Aid for Parents of Children with Disorders of Sex Development**

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The words of parents of children with disorders of sex development inspired me to take on this project. To honor their voices and stay true to the parental perspective I will include selected pieces of their stories throughout this work. Their words have been chosen from a series of interviews conducted by the Sandberg research group at the University of Michigan in their studies investigating the parental experience of making early decisions for their children with DSD. The participating medical center's Institutional Review Board approved all study procedures, including the use of parents' words in this piece.

- Introduction -

**Listening to Parents' Words: A Call to Action**

*We had known that we were pregnant and we decided we wanted to find out if we were having a boy or a girl. We had an ultrasound and the ultrasound lady told us it was a girl. For 20 weeks I had been preparing for a little girl to come into my life. When it came time for delivery I was so excited and nervous. She was finally coming! She was born, and the doctor said, "It's a girl!" I was so happy! But then he said, "... no, wait. It's a boy... Jane, there is something wrong with your baby. We're going to have to take it to perform some tests." We had kind of seen a flash of her in the delivery room, but then she was whisked away to ultrasound, whisked to x-ray, whisked to blood tests, and whisked wherever else before I even got to touch her. At that point, all I remember is knowing that there was something different with our child and they didn't know what we had. I thought I was going to have a healthy baby. My hormones were raging, I was devastated, I was worried, and my whole world came crashing down.*

*When the doctors came back to talk to us, they told us that our child had a disorder of sex development. They said that they weren't sure if we had a boy or a girl. The tests they were running would help them figure out a diagnosis, but they said that the tests could only tell so much. They could not tell us if the baby will feel like a boy or a girl. I didn't know what he was talking about. I asked some questions but I still couldn't wrap my mind around what was happening. I kept using the pronoun "she." For 20 weeks I had been calling her "she," so this was a habit if nothing else. If I didn't use "he" or "she" what pronoun was appropriate? There wasn't one. Every time the doctor would catch me using the pronoun "she" he would cut me off - "Oh, you do understand we have not made the decision about gender yet...do you not?"*

*That I understood. What I didn't understand was why it was taking them so long to figure it out. I mean, what were we supposed to tell our family and friends? They knew I had the baby about a week ago and people had been calling me to see what's going on. But it's a little bit of a personal area of the body so... I don't know. I didn't want to run around with a banner saying that my child has an issue with that part of his body. I didn't know how to talk to them about it. I didn't know what to say. And the doctors didn't offer me any help in that department.*

*Finally the doctors came in one day and explained that this test or another proved that she was a girl. They said that she had a disorder called partial androgen insensitivity syndrome. They tried to explain it to me. A lot of the information they gave us was oral. They did give us some pamphlets and they kind of explained things over and over, but then they were gone and you just have all this stuff to process by yourself. You really want comprehensive information and it's hard; the most stressful thing is just not being presented with clear-cut information. I'd never heard of this before. I had never talked to*

*anybody that had ever experienced it and I kinda felt like... I was the only one ever having to deal with this.*

*I was discharged from the hospital and when I got home I was brave enough to look up her condition online. It was not something I wanted to go to the Internet for, but I needed more information. I think the one thing I've really struggled with is having to pick and chose specific things from different resources to get quality information that is applicable to my child.*

*One day, when I came back to the hospital to visit my child, they told me that there were some decisions that my husband and I would need to make. Some of them, they said, were pretty straight-forward and needed to be done for health reasons. They told me that other decisions would be my choice. In particular, they said there was extra skin on my daughter's genitalia and she would have to have surgery. I could either wait until she got older and have her choose whether or not to do the surgery or I could decide for her now. I don't even know why he gave me the option. It was never even a choice for us. The minute she was born we knew this had to be fixed. We wanted her normal, you know, as normal as possible. We never even discussed not doing the surgery.*

*So then the urologist was called in and, like a ray of sunshine, said "I can fix this." She told us that she could fix her genitals in one surgery and after surgery everything would be fine. We had to wait a bit for her to gain weight, but at one year old she had her first surgery. At one of her follow-up appointments we found out the scaring didn't heal quite right. So then, at two years old, they had to go in and do a little repair. Right now things look good, but I'm still concerned with what's going to happen in the future. Some days I worry about what are things are gonna look like as she gets older. I wonder, will people treat her differently? Will she feel like a boy? Will she like boys? Things are functioning perfect right now, but the surgery has only fixed so much. I guess we'll just have to cross that bridge when we get there.*

*This was the most traumatic thing in my life and I had nowhere to go. I just want to do what is best for my child. I want her to be happy.*

These are the words of many different parents sewn together to tell one complete story: the parental experience of making early medical decisions for their children with disorders of sex development (DSD). As we will see, when it comes to DSD, no two children's situations are exactly the same and no two parents demonstrate the exact same approach to the management of their child's DSD. For these reasons, this vignette should not be taken to represent the exact experience of every parent. Rather, one should read this vignette for its underlying story – namely, parents' shock, stress, uncertainty, thirst for more and better information, and desire to do what is best for their child in the face of all

these hardships. These are the themes that run through most every parent's discussions in some way, shape, or form. Throughout this paper, keep these themes in mind and use them as a way to situate yourself into the parental perspective. For there is power in perspective, and this should not be forgotten.

Disorders of Sex Development (DSD) are defined as "congenital conditions in which development of chromosomal, gonadal, or anatomic sex is atypical." (Lee, Houk, Ahmed, & Hughes, 2006) DSD is an umbrella term that encompasses a multitude of phenotypic and physiologic conditions. Including, for example, aphalia (lacking a penis), vaginal agenesis (lacking a vagina), hypospadias (when the urethral opening is somewhere off the center, tip, of the penis, such as the side of the head, on the shaft, or at the base of the penis), complete androgen insensitivity syndrome (CAIS) (People with CAIS have XY (male-typical) chromosomes but tend to look like a woman with feminine secondary sex characteristics, breasts and a vagina), and Klinefelter syndrome (people with this syndrome have 46'XXY sex chromosomes), just to name a few. Some DSD are diagnosed prenatally or at birth; Some are considered to be severe (and in the case of salt-wasting CAH, potentially life-threatening), but others are much less so; Some people with DSD can appear completely typical, but have underlying physiologic atypicality. And conversely, others can appear phenotypically atypical with no identifiable physiologic etiology. The term DSD is used to signify all of these conditions, but should not be thought of as a diagnosis. Only when physiologic etiology can be identified, such as CAIS, does the person have a diagnosis. Terms like "hypospadias" and "vaginal agenesis" should be thought of as descriptive terms used to identify a specific phenotype. With such a wide variation in

phenotype and physiology presentations of DSD, it is often said that no two individuals (or in the medical context, no two cases) with DSD are exactly alike.

When a child is born with a DSD parents<sup>1</sup> often have to make early decisions regarding the medical and social management of their young child's condition.<sup>2</sup> Parents could be faced with decisions about gender assignment (whether to raise their child as a boy or girl), surgical interventions that are medically necessary (such as the correction of an imperforate anus – i.e. when the rectum has openings into the bladder or urethra. – to reduce the child's risk of potentially life-threatening infections), surgical interventions that are elective (such as clitorplasty to reduce the size of a girl's clitoris), and also whether or not to disclose their child's condition to others, when to do it, and how. These are not always easy decisions. Some decisions are consensus-based: reasonable people tend to agree on one best course of action. Other decisions, however, are value-based: where reasonable people reasonably disagree because what is considered “best” depends on personal values, beliefs and preferences.

There are two primary consensus-based decisions in the medical management of DSD. The first being gender assignment. In cases where the child's gender designation is

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<sup>1</sup> Throughout this paper when I say “parents” let's just assume that I am including all other kinds of legal guardians and caregivers that would be making decisions on the child's behalf.

<sup>2</sup> Throughout this paper when I refer to a “young child” I will use this to designate a child who is unable to participate in decision-making because they lack the capacity to do so. Further discussion of this capacity will occur in Chapter 2. Children who are old enough to participate in decision-making should be provided the resources and encouragement to do so. (American Academy of Pediatrics: Committee on Bioethics, 1995; Lee et al., 2006) However, the incorporation of a child's opinion in this kind of decision-making becomes quite ethically complicated, and for that reason will not be addressed in this paper. Surely components of this discussion like the value of shared decision-making, disclosure, and appropriate information exchange are important for older children and adults making decisions regarding their DSD management.



initially unknown, expert evaluation used in conversation with parental beliefs and intuitions can help determine the most appropriate gender assignment for the child. (Lee et al., 2006) As we will see in Chapter 1, which data is considered relevant to determining the appropriate gender assignment – i.e. gonadal tissue, hormone levels, karyotype, genital appearance, potential gender identity, potential fertility, views of the family, etc. – and how much weight each factor is given, has changed throughout history and remains is still debated today.

There is also general consensus that all interventions necessary to ensure the physical health of the child should be performed. When a child is born with salt-wasting CAH, for example, the child requires daily medication to manage this potentially life-threatening condition. This medication should be provided. Again, it is important to remember that with such variation in DSD conditions and their presentation, two people with the same diagnosis can have very different phenotypic displays. Thus, for one of these individuals an intervention could be considered cosmetic and for the other the same intervention could be medically necessary. These lines are not cut and dry. Each case is unique, and thus, every decision and intervention is case-dependent.

There are a number of value-based decisions parents must make regarding the medical management of their child's DSD. I use the term "value-based" to indicate decisions where there is no clear best option, and when non-intervention is a medically-viable option. Some refer to these value-based interventions as "elective" interventions. Since there is no one right answer, these decisions are largely based upon parents' subjective values. Thus, reasonable people making reasonable decisions tend to disagree. There are many debates that exist within the discourse of DSD about which interventions are the

right interventions, what reasons are the right reasons, or which techniques are the right techniques. Some have questioned if parents are making ethical decisions, others have questioned if parents even have a *right* to make these early value-based decisions. Some of these debates will be outlined in the coming chapters. The important point to take away here is this: In the midst of this controversy, parents have to make complex decisions with their child's healthcare providers during times of high stress and anxiety.

In this paper I will not take sides in the debate that surrounds DSD interventions. First, because there are many voices already in the midst of these conversations that make good (and if not good, than at least valid) arguments both for and against specific value-based interventions, but second – and primarily – because I don't think I have an answer. I find value in some points made by both sides, but these decisions are complex. And I often think that arguments become too simplistic and lack one particularly key ingredient: the parental perspective of making these early decisions.

The debates about early medical intervention for children with DSD rarely reference the parental experience of making these decisions from the *parents'* perspectives. There have been a few qualitative studies that have explored parents' early decision-making experience (For example, Crissman et. al., 2011, Jürgensen, Hampel, Hiort, & Thyen, 2006; Sanders, Carter & Goodacre, 2001), but as these studies have only recently been conducted their findings have not yet made their way into normative DSD rhetoric. Since parents are the ones responsible for making the decisions so frequently debated, their situated perspective needs to be taken seriously.

Having been involved with two different studies of the kind mentioned above, I've heard the words of over 60 parents describe their early experiences of making medical

decisions for their child with DSD. The one thing I've come to appreciate over all others is this: These are real people making real decisions that have real, life-long, consequences for their child. Parents just want to do what is best for their child, and the debates of activists and scholars are far removed from the practical world of what parents are thinking about when making these decisions. As Carl Elliott so perfectly put it:

We have the culture that we have, and we live in the present, not the past. Cultures change, of course, and it is more likely that ours will change if fewer surgeries are done and [people with DSD are] acknowledged openly, but few parents will willingly risk what they believe to be the well-being of their child in order to protest cultural norms. (Elliott, 1998)

Before I spent hours transcribing interviews, coding transcripts and speaking directly to the parents themselves, it was easy to get sucked into academic or philosophical arguments about which parental decisions were the right decisions and which were wrong. In the past three years I've come to see these decisions through the lens of a parent, not just a feminist scholar. In the past three years I've come to understand DSD management in a much more complicated and nuanced way.

What's truly at stake in these discussions is not whether parents are making the *right* or *wrong* decisions (which are evaluated by their ends), but rather, if parents are making *good* or *bad* decisions (which are evaluated by their means). Put another way, it often doesn't matter what decision parents make, what matters is the quality of the process that brings them to that end decision. Parents need holistic, comprehensive, quality, and realistic information about their child's condition, the condition's interventions, and intervention outcomes in order to make informed decisions regarding their child's care. Parents need to be involved in the decision-making process, and they need to determine what their personal beliefs and values are so that they can work with their child's

healthcare providers to make decisions that best match their subjective preferences. If parents have this information and have participated in this way, they have made a good decision. Decisions of this kind are known as “informed shared decisions” and they are acquired through the processes of informed decision-making (often referred to as informed consent) and shared decision-making – i.e. “informed shared decision-making.”

As the vignette above highlights, this is not always the case. Sometimes parents don't even conceptualize that there are decisions that need to be made – *“I don't even know why he gave me the option.”* Sometimes parents lack understanding of diagnostic tools and clinical findings – *“Finally the doctors came in one day and explained that this test or another proved that she was a girl.”* Sometimes parents are lacking realistic outcomes – *“Then the urologist was called in and, like a ray of sunshine, said “I can fix this.” She told us that she could fix her genitals in one surgery and after surgery everything would be fine.”* And don't forget that parents are making these decisions during times of high stress and anxiety – *“My hormones were raging, I was devastated, I was worried, and my whole world came crashing down... This was the most traumatic thing in my life and I had nowhere to go.”* Parents' words have illuminated gaps in the informed shared decision-making process regarding the medical management of their child's DSD. And in their words I have heard a call to action: *“You really want comprehensive information and it's hard; the most stressful thing is just not being presented with clear-cut information”* – That is, “help us acquire the information we need to make good decisions for our child, and help us through this decision-making process so that we can do what is (“clearly”) best for our child.” The discussion that follows is a response to this call.

In particular, I will address the development of a decision-aid for parents making early medical decisions for their children with DSD. Decision-aids are tools often used to facilitate the shared decision-making process and have been found to enhance decision-making in a number of meaningful ways. Including, but not limited to, patients' a.) improved knowledge of the options; b.) more accurate expectations of possible benefits and harms; c.) increased participate in decision making; d.) increased patient-practitioner communication; and e.) increased selection of choices that are more consistent with their informed values (when the decision-aid includes values-clarification exercises.(D Stacey et al., 2011) Patient decision-aids do *not* advise people to choose one option over another. And they do *not* replace counseling from healthcare practitioners. Rather, they help people make value-based decisions where the best choice involves matching personal values, situational beliefs and the features that matter most to a person with the option that has these features.(Steering Committee, 2005) To date, over 500 decision-aids have been created for a wide variety of conditions. None of them are DSD. A decision-aid specifically tailored to address the early decisions parents must make, can help ensure that parents are making the *best* decisions for their children. After all, as the parent above said, "*I just want to do what is best for my child.*" A decision-aid is an answer to that call.

#### A NOTE ON TERMINOLOGY

In the past, the terms "hermaphrodite" and "pseudohermaphrodite" or "intersex" have all been used in both the medical and social areas to describe people with these kinds of conditions I described above as disorders of sex development (DSD). Throughout this paper, I will use the term "disorder of sex development." The reason is two-fold.

First, the terms “hermaphrodite,” “pseudohermaphrodite,” and “intersex,” each define the atypicality of sexual development in vague and different ways. Parents and practitioners alike found them to be confusing and difficult to utilize in the clinical setting. Additionally, some parents and people with DSD have resisted the terms “hermaphrodite” or “intersex,” claiming that they seem sexualizing, they act to eroticize an issue that is instead a medical or biological one, and they imply no clear gender identity while simultaneously placing an intersex identity onto the person with DSD.(Dreger & Herndon, 2009) Understood in these ways, the terms presented barriers to conversations, collaboration, and improvements in care because they were clinically unclear(Lee et al., 2006) and many associated the terms with “radical gender activists who advocated deferral of sex assignment and opposed early genital or gonadal surgeries.”(Feder & Karkazis, 2008)

In 2005 at the International Consensus Conference on Intersex, organized by the Lawson Wilkins Pediatric Endocrine Society (currently the Pediatric Endocrine Society) and the European Society for Pediatric Endocrinology, participants proposed

TABLE 1 Proposed Revised Nomenclature	
Previous	Proposed
Intersex	DSD
Male pseudohermaphrodite, undervirilization of an XY male, and undermasculinization of an XY male	46,XY DSD
Female pseudohermaphrodite, overvirilization of an XX female, and masculinization of an XX female	46,XX DSD
True hermaphrodite	Ovotesticular DSD
XX male or XX sex reversal	46,XX testicular DSD
XY sex reversal	46,XY complete gonadal dysgenesis

(Lee et al., 2006)

the term “Disorders of Sex Development” (DSD) to replace the terms “hermaphrodite,” “pseudohermaphrodite,” and “intersex.” The proposed terminology was meant to address the previous terms’ obscurity by breaking down the umbrella term – disorder of sex development – into clinically relevant sub-categories. The sub-categories “46,XY DSD,”

“46,XX DSD,” “ovotesticular DSD,” “46,XX testicular DSD,” and “46,XY complete gonadal dysgenesis” indicate the molecular genetic aspects of sex development while simultaneously leaving space for the spectrum of phenotypic variation present in DSD.(Lee et al., 2006) The new terminology also acts to address the potentially pejorative nature of the previous terms by expressing “person first” ideology. An ideology that acknowledges that the *personhood* of an individual should be taken into account before any other reference.(Maio, 2001) This, in effect, emphasizes the fact that (at least in the medical context) one cannot *be* intersex; one *has* a DSD – and thanks to the nomenclature of sub-categories: one has this specific *kind* of DSD.

Not everyone has taken up the new terminology with open arms. Some advocacy groups continue to use the term “intersex” because they object to the use of the term “disorders” which they believe effectively acts to medicalize conditions that they believe should be outside the medical domain.(Feder, 2009) This leads us to the second reason that I use the term DSD throughout this paper: Since the goal of this paper is to address decision-making in the medical realm and the medical realm uses the term DSD, then it is only fitting to use the rhetoric of the medical domain. However, I do have some reservations about the term. As mentioned in the introduction, the word “disorder” implies that a DSD is something you must treat.(Feder, 2009) While some medical interventions for DSD are medically necessary for the physical health of the child (and for that reason the term “disorder” is quite fitting), others are not. When it comes to these *other* interventions, like cosmetic surgery on external genitalia, parents need to know that the decision not to intervene is equally open to them. But in a culture and practice where the state of being “ill” – i.e. having a disease or *disorder* – is in itself undesirable there is an obligation to get

well.(Ablon, 1981) Similarly, when Western biomedicine has access to the technology used to “fix” the “problem,” there is an obligation to use it. This is known as the “technological imperative.”(Davis-Floyd, 2003) This parent’s words reflect this pressure to intervene that follows a diagnosis:

*Did she even need to have that [surgery] in the first place? Should we have just left it alone?... It seems like with doctors, it’s such a, like they just want to fix it and diagnose.*

The distinction between interventions that are medically necessary and those that are elective is not always clear to parents and implications of the term “disorder” do not help distinguish between the two. When using the term “DSD” extra vigilance needs to be taken to make sure parents understand which aspects of their child’s condition can and should be treated with specific interventions, and which aspects of their child’s condition.



- Chapter 1 -

**The History of Disorders of Sex Development  
and Where we are Today**

*When the doctors came back to talk to us, they told us that our child had a disorder of sex development. They said that they weren't sure if we had a boy or a girl. The tests they were running would help them figure out a diagnosis, but they said that the tests could only tell so much. They could not tell us if the baby will feel like a boy or a girl. I didn't know what he was talking about. I asked some questions but I still couldn't wrap my mind around what was happening. I kept using the pronoun "she." For 20 weeks I had been calling her "she," so this was a habit if nothing else. If I didn't use "he" or "she" what pronoun was appropriate? There wasn't one. Every time the doctor would catch me using the pronoun "she" he would cut me off - "Oh, you do understand we have not made the decision about gender yet...do you not?"*

Susan Graseck said, "Explore the past to understand the present and shape the future." In order to accurately place yourself in the middle of conversations regarding DSD care today, you must understand how these current conversations came to be. You must understand the foundations. You must understand the history. This chapter will go through the medicalization of people with DSD and the way conceptualizations and interventions have changed through time, ending with the state of care today. Because the history of DSD is largely shaped by definitions and redefinitions in a multitude of arenas, I will often refer to people with DSD as "individuals with atypical somatic sex development." This more neutral terminology avoids definitional conflict with the terms used to refer to people with these kinds of conditions at a specific point in history or from a specific perspective. This history will provide the backdrop for all discussions that will follow in this paper.

TACKING DOWN DEFINITIONS AND INCIDENCE RATES

In 1945 a human anthropometric model named Norma – pun definitely intended – was created based upon composite measurements of thousand of young American

women's bodies. Norma became a plastic celebrity of sorts. Characterized as the "ideal young woman" of the time, everyone wanted to see how they "measured up" – again, pun intended – to the average American woman. Under the tag line, "Are you Norma, Typical Woman?" a search to find the woman in Ohio who most resembled Norma began. Women were instructed on how to measure their bodies and within ten days nearly 4,000 women had entered. Here's the kicker: of the 4,000 women who entered, less than one percent of women came close to Norma's proportions!(Urla & Swedlund, 1995) As it turns out, the average composite of the averages of women isn't normative at all! As Shapiro explains, "The average American figure approaches a kind of perfection of bodily form and proportion," the average of all averages – i.e. Norma – is actually "excessively rare."(Shapiro, 1945)

Today, the Norma case serves as a reminder that what is "normal" and what is "average" are two different things – especially when you are combining multiple factors into one composite. Norma may be average, but she sure wasn't typical. With all the combinations of characteristics that make up bodies in the world, it's difficult to determine the boundary between "typical" and "atypical" even when we have descriptive averages of bodily forms. This ambiguity in defining normalcy has proven particularly troublesome in relation to DSD and human sexual anatomy. There are definitely norms for human sexual anatomy: we expect men to have male-specific internal sex organs, a penis and two testicles that look something like "this." And we expect women to have female-specific internal sex organs; a vagina and clitoris that look something like "that." However, the "this" and "that" that exist in the realm of "average," have great breadth and diversity for typical men and women.

For example, a woman's vagina can be short, long, deep or shallow. Her labia can be flat or ruffled. Her clitoris can be small or large. A man's penis can be straight, curved, short or long. It can be circumcised or uncircumcised. And his testicles can be big or small, hang low or be relatively close to the body. As long as these phenotypic appearances are not due to an underlying pathology, a wide breadth of sexual anatomy is considered to be typical.

With all this diversity, it can be very difficult to define the point when human sexual anatomy moves into the realm of atypical. What combination of short, long, big, or small makes someone atypical? What combination of these physical factors should put up a red flag for physicians to further investigate the etiology of a particular appearance, or suspect that the person's genital appearance is due to pathophysiology? In the case of DSD, sometimes these questions are easily answered: When a child is born with *ambiguous genitalia* – that is, some combination of male-typical and female-typical anatomy – the red flag burns bright. Other times, however, identifying the realm of the atypical is not that easy. Take, for example, an adult woman who is otherwise healthy but has a very small vagina – so small that she is unable to use a tampon. Her vagina is small, yes. But is it so small that it should be considered pathological? One can use statistical criteria (e.g.  $<-2SD$  or  $>+2SD$ ) or functional criteria (can the person complete a biological function) to try and answer this question, but until the boundaries of “typical” versus “atypical” are established there will always be controversy. And because these boundaries are largely subjective, one could argue that to some extent their establishment will always be in the eye of the beholder. This is the first of a few reasons why it is difficult to provide definite incidence rate of individuals living with a DSD.

Another reason that determining an accurate number of individuals living with DSD is difficult to achieve is that the incidences of various conditions included under the umbrella term “DSD” vary between populations and even geographic locations. Some DSD, like ovotesticular DSD, are fairly rare, occurring in 1/10-20,000 live births. Other DSD, like hypospadias, are fairly common and are estimated to affect one in every 150 boys.(Joosten et al., 2008) Also, populations can vary greatly in incidence rates for the same condition. For example, the calculated incidence for late-onset adrenal hyperplasia is 37/1,000 among Ashkenazi Jews, 19/1,000 among Hispanics, 3/1,000 among Italians, and 0.01/1,000 among mixed Caucasian populations.(Arnaut, 1992; Eldar-Geva et al., 1990; Newfield & New, 1997; White, New, & DuPont, 1987) Without taking this underlying diversity in subgroups into account, an overall DSD incidence rate can actually be quite deceiving in relation to the entire population.

There is also some debate over which conditions should be included under the DSD umbrella term. In 2000, Blackless et. al. stated that these kinds of conditions occur in 1.7% of the population, while an estimated 1.62% of the population may be subject to genital surgery as an intervention for their DSD.(Blackless et al., 2000) However, in 2002 Leonard Sax issued a response paper to this study stating that the 1.7% estimate was deceptively large because it includes conditions that “most clinicians do not recognize as intersex, such as Klinefelter syndrome, Turner syndrome and late-onset congenital adrenal hyperplasia (LOCAH).”(Sax, 2002) Sax argued that 47’XXY men with Klinefelter Syndrome and 45’X0 women with Turner Syndrome should not be considered to have a DSD because they generally do not have ambiguous genitalia. However, using the definition of DSD provided in the Consensus statement – “congenital conditions in which development of

chromosomal, gonadal, or anatomic sex is atypical” (Lee et al., 2006) – these two conditions are clearly included due to chromosomal atypicality, hypogonadism, and infertility.

Similarly, because the genitalia of LOCAH children are normal at birth, Sax believes that they too should not be considered to have a DSD. Without LOCAH, the number of DSD-related surgeries falls to 0.08% of the population, or between one and two in every one thousand births. After Sax excluded conditions that he argued should not be considered DSDs, his calculated frequency fell to 0.018% – almost 100 times lower than the estimate provided by Blackless et. al.. The true frequency of DSD and their interventions probably falls somewhere in between these two approximations that essentially bookend the inclusion vs. exclusion spectrum. (Blackless et al., 2000)

As we will see, history and culture also determine the identification and classification of people with DSD, causing there to be a variation in the criteria used to determine what constitutes “male” and “female” and the significance of these differences. The knowledge of human physiology of the time, the medical technology, and larger socio-cultural context all play a role in defining these criteria. For these reasons, the stats on DSD change based on who is counting at a given time and place. (Hester, 2004) While it is true that the intention of identifying incidence rates is often aimed at academic rather than therapeutic ends, one must examine the ways in which definitions used to acquire incidence data affect those therapeutic ends. The distinction is never as clean as one might assume *a priori*.

For example, imagine a woman with LOCAH, which causes her clitoris to grow so significantly that she must make decisions about potential cosmetic genital surgery. If she could benefit from the care provided by DSD multidisciplinary teams, why should she be

excluded from this care? Similarly, some physicians have argued that when hypospadias – a condition where a boy’s urethral opening is somewhere off the tip of his penis (such as the base, along the shaft or off-center on the tip) – has no identifiable etiology, it should be considered “isolated hypospadias” and not a DSD. Compared to other, more severe, forms of DSD, having a urethra opening slightly off the tip of the penis seems of little worry to practitioners. But when it comes to genital atypicality, we know that parents’ stress is not associated with the severity of the condition (DeMaso et al., 1991; Tak & McCubbin, 2002; Youngblut & Shiao, 1992), and we therefore must remember that something that seems “simple” to practitioners may not feel the same way to parents. When individuals in both of these scenarios would benefit from being cared for by a trained, multidisciplinary team that is sensitive to the physical and social complexity of DSD, why should they be denied this care? Simply because we are having word wars over definitions does not seem to justify excluding these populations from our conversation and interventions. It’s important to remember that these conditions affect real people and their families. It is estimated that in the US and Europe alone, there are tens of thousands of children born with DSD each year, with an average of between five and ten surgeries performed per day throughout the Western/European world. (Hester, 2004) This frequency is greater than that of cystic fibrosis (1 in 2,500 “Caucasian” births) (The Cystic Fibrosis Foundation, 2012) or Albinism (1 in 17,000 births) (The National Organization for Albinism and Hypopigmentation, 2012) – conditions that receive comparatively much greater public awareness. (Hester, 2004)

## CONTEXT &amp; CONCEPTUALIZATION OF DSD

Definitions aside, the percentage of people living with DSD in the population has not been stagnant throughout history. In the past, certain health-related aspects of DSD would have been fatal before the development of specific medical knowledge and interventions used to address them. Additionally, preliminary studies have shown that changing environmental factors may play a role in the number of individuals living with DSD: Increased prenatal fetal exposure to environmental hazards known as “endocrine disruptors”<sup>3</sup> affect fetal sexual development and sexual differentiation (Jacobsen, Christiansen, Boberg, Nellemann, & Hass, 2010) – including ‘idiopathic’ partial androgen insensitivity syndrome (PAIS)-like phenotype (Gaspari, Paris, Philibert, et al., 2011) and an increased risk for male urogenital malformations (Fernandez et al., 2007; Gaspari, Paris, Jandel, et al., 2011) including hypospadias (Morales-Suárez-Varela et al., 2011). While these studies show a correlation between exposure to endocrine disruptors and atypical sexual development, the effects of these disruptors may not cause bodies to cross the definitional threshold into DSD from the generally accepted variations of “normal.”

These statistical considerations are important to keep in mind as they affect the likelihood of exposure of these conditions to practitioners and larger social awareness in the public realm. However, more central to the issues at hand is not the question, “How

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<sup>3</sup> As defined by the National Institute of Environmental Health Sciences (NIEHS), “Endocrine disruptors are chemicals that may interfere with the body’s endocrine system and produce adverse developmental, reproductive, neurological, and immune effects in both humans and wildlife.” Wide ranges of substances, both natural and man-made, are thought to cause endocrine disruption. These substances include “pharmaceuticals, dioxin and dioxin-like compounds, polychlorinated biphenyls, DDT and other pesticides, and plasticizers such as bisphenol” and they can exist in the environment as well as many everyday products such as “plastic bottles, metal food cans, detergents, flame retardants, food, toys, cosmetics, and pesticides.” (NIEHS, 2012)

many people with DSD are there in the population?” but rather, “How have people with DSD been conceptualized and treated by the larger population?” As we will see, DSD care and management has changed through time depending on multiple factors. The status of the medical profession itself, medical knowledge (including, for example, understanding of hormones, genetics, and sexual development), advances in medical technology (including, for example, surgery and surgical techniques, and anesthesia), and societal conceptualizations of sex, gender, and sexuality all influence the care and management of people with DSD. If we take a quick walk through time we see just how dependent the conceptualization of DSD is on all of these other factors.

#### A WALK THROUGH TIME

Until the mid-nineteenth century people with atypical genitalia tended simply to “blend in with the general population, living their lives as unremarkable boys, girls, men, and women.” (Dreger & Herndon, 2009) When doctors began to enter the professional arena as people with authoritative knowledge, however, those people living with a DSD also entered the medical realm. When individuals with DSD came into a doctor’s practice for one reason or another – presumably to receive a diagnosis, relieve discomfort or pain, address sex dysfunction, fertility problems, and/or other issues faced by people with DSD – doctors became intrigued by their “unique” bodies and wanted to share their findings with the medical community. (Dreger, 1998) In the mid-nineteenth century people with DSD started making more appearances in medical and legal journals and textbooks. Doctors often wrote a detailed description of the ambiguous physical condition of the patient, their marital, parental, racial, and class status, and the doctor’s opinion of the person’s true



sex.(Reis, 2005) These nineteenth century *descriptive* publications were written about people with DSD mostly to show their story to the medical world, not to prescribe interventions. To the extent that interventions were done, they were performed in attempt to classify people as either male or female based upon their “true sex.”(Dreger, 1998; Karkazis, 2008)

The methodologies used to determine an individual with DSD’s true sex evolved over time. In 1876, the German pathologist Kelbs was the first to suggest that gonadal tissue be the sufficient factor to determine an individual with DSD’s true sex. Kelbs used the taxonomy of “true hermaphrodites” and “pseudo-hermaphrodites” to distinguish between those individuals who had *both* testicular and ovarian tissue from those people with gonadal tissue that did not correspond with their phenotypic appearance.(Karkazis, 2008) A male pseudo-hermaphrodite, for example, would have been someone who appeared to be a woman in every respect with feminine phenotype (breasts, a vagina, etc.), but also had testicular tissue. No matter what the person looked like – small penis, larger clitoris, some combination of both – a person’s true sex was determined by their gonadal tissues. This idea didn’t take off until 1896 when British experts George Blacker, and T.W.P. Lawrence professed the simplicity of this gonadal approach in determining the “true sex.”<sup>4</sup>(Dreger, 1999) Testicular tissue was found in men, and ovarian tissue was found in women. It was that simple. European and American medical professionals soon rallied behind the idea. The gonadal approach to sex determination cleanly placed people into neat groups: Man, woman, and true hermaphrodite. Thus establishing clean definitional boundaries.

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<sup>4</sup> This period of understanding was coined, “the age of the gonads” by Alice Dreger in her book, “Intersex in the Age of Ethics.”

These neat distinctions did not last long. Advances in surgical techniques including anesthesia and antiseptics, the medicalization of the childbirth, new understandings of sex differentiation and embryology, and in particular the discovery of sex hormones during the first half of the 20<sup>th</sup> century reshaped what was relevant to the classification of individuals with DSD.(Dreger, 1999; Karkazis, 2008; Matta, 2005)

In 1891, the French physiologist Charles Edouard Brown-Sequard addressed his colleagues at the Society of Biology in Paris to present the findings of his latest experiment. Brown-Sequard had been giving himself injections of crushed guinea pig and dog testicles and found that they left him feeling spry and rejuvenated. While admittedly disgusting in method, Brown-Sequard's findings made major scientific advances in the knowledge of animal physiology. He discovered that specific organs secreted substances that he called "internal secretions" that affected other aspects of the body.(Borell, 1976)

At the beginning of the twentieth century Brown-Sequard's theory of "internal secretions" was refined by the British physiologist Ernest H. Starling. Starling introduced the concept of hormones, saying that "these chemical messengers or 'hormones' as we may call them, have to be carried from the organ where they are produced to the organ which they affect, by means of the blood."(Starling, 1905) The chemical messengers believed to originate from the gonads were designated "sex hormones." Male sex hormones were secreted by the testes and controlled "dependent male characteristics." Female sex hormones were created by the ovaries and controlled "dependent female characteristics."(Oudshoorn, 1990) Brown-Sequard's unmatched dedication to science with self-administered crushed testicle injections and Starling's concept of the hormone directly

challenged the gonadal definition of sex: it wasn't *just* the gonads that defined a person's sex, gonads *and* hormones were sex-specific indicators.

In 1915 physician William Blair Bell found these new discoveries in endocrinology quite moving. Published in the Liverpool Medico-Cirurgical Journal, he stated, "Since it is now possible to demonstrate the fact that the psychical and physical attributes of sex are not necessarily dependent on the gonads, I think that each case should be considered as a whole; that is to say, the sex should be determined by the obvious predominance of characteristics, especially the secondary, and not by the non-functional sex-glands alone, for this is neither scientific nor just." (Bell, 1915) Bell's position reflects previous ideology in that it continued to uphold clear distinctions between the two sexes and support for a *true sex* for every individual. More importantly, by taking secondary sex characteristics into account, Bell introduced a more complex and nuanced understanding of what can be used to define a males and females, men and women.

In 1917 the term "intersexual" was born out of these new discoveries in endocrinology. German scientist Richard Goldschmidt's research on hormone action showed that sexual differentiation into one body type or the other was mediated by hormones and controlled by hormone-producing tissues. When hormone action went awry, sexual development could produce individuals that displayed both male and female somatic characteristics. Goldschmidt called these individuals "intersex" as a replacement for the term "hermaphrodite." This term soon entered the medical arena where clinicians began using it to refer to people with various somatic forms of sex atypicality. (Goldschmidt, 1917 in Dreger, 1998 and Karkazis, 2008)

The study of individuals with atypical somatic sex development has long been conducted with the assumption that understanding atypical sexual differentiation (from both a physical and psychological point of view) will both enhance understanding of more typical sexual differentiation (Zucker, 2002) and reinforce the normative definitions and boundaries of typical sexual differentiation (Dreger, 1998; Hester, 2004; Karkazis, 2008; Matta, 2005). Social scientists and historians have pointed out that – as with all medicine, throughout history – interventions for people with DSD have largely been influenced by the broader sociocultural arena. In particular, a number of historians have pointed out the connections between the history of interventions for people with DSD and the history of homosexuality: (Dreger, 1998; Karkazis, 2008; Matta, 2005; Reis, 2005) “The medicalization of hermaphroditism, already concerned with gender boundaries,” Matta writes, “became entangled in the medical profession’s redefinition of homosexuality as a matter of sexual object choice, and therefore fell doubly victim to the perceived need to define appropriate sexual and social behaviors.” (Matta, 2005) The late 19<sup>th</sup> century push for gonadal tissue to be the defining feature of one’s true sex is an example of how medicine was used to help clean up blurring social lines by neatly placing individuals into definite groups. (Dreger, 1998)

As this section revealed, the interventions physicians used to classify people changed through time in relation to accessible medical technology, physiological understanding and larger sociocultural influences. Before the middle of the twentieth century these interventions were mostly meant to sort people into the distinct groups formed by normative dichotomies – such as boy vs. girl, man vs. woman, heterosexual vs.

homosexual. It wasn't until the mid-twentieth century that hormonal and surgical interventions became the standard of care used to *treat* people with DSD.

#### THE BIRTH OF SURGICAL INTERVENTION AND MONEY'S PARADIGM

Surgical interventions for DSD have actually been around for quite some time. The first report of corrective genital surgery was written in 1852 by Samuel D. Gross and published in *The American Journal of Medical Sciences*. In 1849 he performed this first surgery on a three-year-old girl whose parents became concerned when she started to show "boyish" interests by playing sports rather than playing with her dolls. Upon the parents' request, Gross examined the girl and determined that she had neither a penis nor a vagina, but a clitoris and a small indentation covered by a membrane and testes. Gross surgically removed her testes and was pleased upon follow-up examination to find that the girl had returned to her feminine interests.(Gross, 1852) Surgical interventions – including those that were considered "corrective" and used on children – did exist as early as the 1850's However, these surgeries were rare.(Dreger & Herndon, 2009; Matta, 2005; Reis, 2005)

Surgical "normalization" became a standard tool used in the medical management of individuals with atypical somatic sex development in the 1950's after the psychologist John Money, working in collaboration with a multidisciplinary team at John's Hopkins University, developed what came to be known as the "optimum gender policy."(Dreger & Herndon, 2009; Gross, 1852) This policy evolved from the findings of a study of 105 hermaphrodites found that only 5 of the 105 patients had a gender role/identity

(psychosexual orientation)<sup>5</sup> and/or sexual orientation that Money et. al. considered ambiguous or discordant with the patient's gender of rearing. Thus, Money et. al. concluded that "the sex of assignment and rearing is consistently and conspicuously a more reliable prognosticator of a hermaphrodite's gender role/identity and sexual orientation than the chromosomal sex, the gonadal sex, the hormonal sex, the accessory internal reproductive morphology, or the ambiguous morphology of the external genitalia."(John Money, Hampson, & Hampson, 1957) Money and his colleagues did not say that social environment and rearing were the *only* factors that played a role in the development of the child's gender role, rather that they played a role.<sup>6</sup> Money and his colleagues were the first clinicians to incorporate psychological factors into DSD care. The optimum gender policy represented a departure from early works on hermaphroditism and intersex that "overwhelmingly focused on usually one aspect of the body, and not the psyche for gender assignment."(Karkazis, 2008)

So what does this have to do with surgery, you ask? Under this paradigm, Money professed that all children were psychologically neutral from birth until the age of eighteen months. As such, all/any interventions needed to ensure a sound psychosexual identity and orientation in the child should be done "before the establishment of gender roles gets far

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<sup>5</sup> Money used the term gender role/identity to indicate a person's internal identification and external display of a given gender. Today the terms are thought of as two of three distinct components that comprise an individual's psychosexual development: As defined by the Consensus Statement, "gender identity" refers to a person's self-representation as male or female (with the caveat that some individuals may not identify exclusively with either); "gender role" (sex-typical behaviors) describes the psychological characteristics that are sexually dimorphic within the general population, such as toy preferences and physical aggression; and the third component being "sexual orientation" refers to the direction(s) of erotic interest (heterosexual, bisexual, homosexual) and includes behavior, fantasies, and attractions.(Lee et al., 2006)

<sup>6</sup> Money et. al. recognized the role prenatal exposure to hormones could have on a child's gender identity.(John Money & Ehrhardt, 1972)

advanced.”(J. Money, Hampson, & Hampson, 1955) Money believed that when a child was born with a DSD, a full medical workup should be done by a multidisciplinary team to determine the best gender of rearing – that is, determine which gender would ultimately be most believable and satisfying for the individual after interventions to “fix” any ambiguity were completed. After the best course of action was determined, the child should then be made into either a boy or a girl via surgically altering the appearance of their genitals to match the given gender assignment as soon as possible.<sup>7</sup>

According to Money, the reasoning for this early surgical intervention was two-fold: First, Money felt that the child was more likely to develop a proper gender identity/role with genitals that matched the assigned gender. Second, Money believed that an important factor in the success of these interventions – i.e. a healthy, happy child with a stable gender identity and actor of appropriate gender roles – was the parents’ commitment to raising their child as the assigned gender. Money believed that early surgery to “normalize” the genitals was necessary to the development of appropriate gender identity in the child and also to avoid any hesitation parents may have toward raising their child in the assigned gender due to the child’s “contradictory” genitalia. Since Money believed that gender role/identity is, in part, learned through socialization via the parents, it followed that how parents acted around their child would directly influence the child’s acceptance of their gender assignment.

Money’s treatment policy correctly reflects the fact that socialization is embodied. The gender assignment (when young) and the gender identity and gender role (when

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<sup>7</sup> Additionally, hormonal interventions were sometimes implemented around the time puberty should emerge to insure the appropriate development of secondary sex characteristics.

older) of others gives people a framework for understanding and interacting with the person. In a study, brought to my attention in Kenneth Zucker's work, a group of researchers showed that the perceptions of a toddler's behavior were affected by labeling as a male, a female, or a hermaphrodite.(Zucker, 2002)

In this study several hundred health professional trainees (e.g. medical and psychology students) viewed an 8-minute videotape of a 22-month-old infant engaged in various activities. Every 15 seconds, the trainees were asked to rate the toddler's last activity as masculine, feminine or neutral. Findings showed that activities were more likely to be rated as masculine than feminine when the toddler was labeled a male, whereas the converse occurred when the toddler was labeled a female. When the toddler was labeled a hermaphrodite, a similar proportion of the activities were rated as masculine and feminine.(Delk, Madden, Livingston, & Ryan, 1986)

Implicit understanding of what it means to be a boy or a girl influences our subconscious and conscious interactions with an individual. Money was correct when he said that gender role is "built cumulatively through experience encountered and translated – through casual and unplanned learning, though explicit instruction and inculcation, and through spontaneous putting of two and two together..." and that once it was learned, it was not easily modifiable.(J. Money et al., 1955) Parents teach their children how to "do"<sup>8</sup> a gender by interacting with them. To support proper gender acquisition in the child, parents have to fully commit themselves to the gender assignment of their child in order to consciously and subconsciously instill this embodiment onto their child.

Money recognized that the psychological stress parents often experience due to their child's condition could prohibit the best outcomes for the child. In order to avoid psychological confusion on the parents' part – and as the child grew, on the child's part as

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<sup>8</sup> "Gender is not something we are born with, and not something we have, but something we do."(West & Zimmerman, 1987) These are the famous words of Candace West and Don Zimmerman in their piece, "Doing Gender," where they explain how gender is embodied and is something that we act out in every instant.



well – physicians often did not inform parents and patients of everything they knew about the DSD if they thought it would complicate or confuse parents’ and persons’ with DSD conceptualization of gender.(Dreger, 1999) An example of this was provided by Katrina Karkazis: “When it came to genital “normalizing” surgery,” she said, “Money suggested that parents be told that their child was either a boy or a girl as soon as possible and that the child’s sex organs were undifferentiated or “sexually unfinished” but would be “finished” through the genital surgery.”(Money, 1955; Money & Ehrhardt, 1972 in Karkazis, 2008) While his intentions may have been – what he believed to be – in the child’s and parents’ best interests, activists, patient advocates, and bioethicists alike have recently challenged this lack of disclosure and information exchange used under the optimum gender policy.

Today Money’s optimum gender policy is faced with criticism in both the scientific and public arenas. In the heat of these debates, it’s easy to criticize specific aspects of what the policy does and does not advocate. Far less attention is given to the groundbreaking contributions Money’s work brought to the medical management of DSD: First, Money’s paradigm essentially acted as the first guideline to best practice for the care of individuals with DSD. His protocol provided clinicians with ways to intervene to help people with these conditions for which previously they had no guidance.<sup>9</sup> The widespread publication and references to Money’s clinical work and research across a plethora of disciplines, specialties, and institutions made Money’s paradigm well known and easily accessible. Since the development of these guidelines to clinical care, Money’s work has been cited in nearly every clinical article on the medical management of people with DSD.(Karkazis,

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<sup>9</sup> This is not to say that all medial practitioners adhered to the practices put forth by the optimum gender policy in every instance they cared for an individual with DSD. The point here is that Money’s paradigm provided practitioners with evidence-based guidance for making these decisions.

2008) Second, Money and his team also introduced the idea of using a multidisciplinary team to better address the many aspects of DSD management.(Karkazis, 2008) This approach to care is endorsed by current suggestions for care of individuals with DSD. Most importantly, Money's approach incorporates psychological considerations of both the parents and child with DSD. In many ways, the optimum gender policy represented a departure from, and an improvement on, earlier work on individuals with atypical somatic sex development that it took the individual's potential psychosexual orientation into account. The same parties that challenge other aspects of the optimum gender policy support these aspects of DSD care today. They just fail to point out these successes with the same ferocity they use to criticize its failures.

#### PARADIGM SHIFT AND DSD MANAGEMENT TODAY

Since the beginning of the 1990's, some of the interventions used in Money's optimum gender policy entered a state of great debate and controversy. Changing social tides, new scientific findings, and re-evaluation of ethical decision-making in the medical field began to challenge some of the standards professed and utilized under Money's paradigm. These controversial discussions ultimately lead to a redefinition of what quality care for individuals with DSD would look like and entail. The transformation from Money's paradigm to birth of the new paradigm under development today will be outlined in this section.

Better scientific understanding of gender, sexuality and somatic sex development has played an important role in the current paradigm shift. Clinicians and scientists have been persuaded that complex developmental processes affect these factors – including both

prenatal and postnatal hormone levels, genetic influences, and postnatal environmental and psychological influences.(Yang, Baskin, & DiSandro, 2010) Most notable to changes in DSD care are studies about the development of gender identity: Multiple studies support what is known as “Brain Organization Theory” which suggests that steroid hormones during fetal development permanently organize the brain in gender-specific ways with respect to sexual orientation, cognition, temperament, and interests that are considered “feminine” or “masculine.”(Hines, 2004, 2011; Institute of Medicine, 2001) With the findings presented above, clinicians today take the potential effects prenatal hormone levels could have on a child’s gender identity into account when making decisions about gender assignment(Accord Alliance, 2012), while simultaneously keeping in mind the separability of sex-typical behavior, sexual orientation, and gender identity. (Lee et al., 2006) i.e. homosexual orientation (relative to sex of rearing) or strong cross-sex interest in an individual with DSD is not an indication of incorrect gender assignment.

In addition to those advances in scientific understanding of DSD, there have also been recent shifts in the social conceptualization of what it means to be a person with DSD. Thanks to a number of identity liberal rights movements, the last twenty years have been marked by tides of activism that have most definitely influenced the care individuals with DSD receive. According to historian and activist Alice Dreger and her colleague April Herndon, the LGBT<sup>10</sup> rights movement, “which brought about positive changes in social attitudes toward queer-identified people,” has lead to positive changes in social attitudes toward those people living with atypical somatic sex development (Dreger & Herndon, 2009) – people with what some call “queer bodies.”(Holmes, 2000) Most notably, the LGBT

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<sup>10</sup> Lesbian, Gay, Bisexual, Transsexual (LGBT)

movement has challenged the set of normative binaries that exist in society. By suggesting that sex (male vs. female), gender (boy vs. girl, man vs. woman), and sexuality (heterosexual vs. homosexual) should be thought of as spectrums rather than as binaries, the movement has made definitional and social space for those people who don't cleanly fit into one end or the other of these binaries.

The disability rights movement has helped deconstruct another social binary, namely, able-bodied vs. disabled. This movement has pushed "person-first" ideology where one should think of diagnosis or conditions as a part of a person, but not the defining characteristic of the person as a whole. Put another way, diagnoses or conditions should not be used to indicate what *type* or *kind* of person one is. In the past, activists, scholars, and journalists sometimes used the term "intersexuals" to describe individuals with atypical somatic sex development, but, as professed by the disability rights movement, this term has "largely fallen out of favor because it can be essentializing and dehumanizing to equate people with one aspect of their physicality." (Dreger & Herndon, 2009) Instead, today when a child is born with atypical somatic sex development, that individual is called a "person *with* a DSD" or sometimes "person with intersex." Some advocates and advocacy groups continue to use the term "intersex" as a form of identity, but this identity is not one automatically placed onto children born with these conditions. Should the child grow up and choose to take on this identity that would be his or her choice.

The intersex rights movement, informed by "principles of feminism (particularly the right to speak for oneself and critiques of sexism), gay and lesbian rights (particularly critiques of heterosexism and homophobia), and patients' rights (especially regarding autonomy, informed consent, and truth telling)" (Chase, 1998; Dreger & Herndon, 2009),

was started by activist individuals with DSD (often referred to as “patient advocates” or “intersex activists”). Cheryl Chase, founder of ISNA and now living as Bo Laurent, along with other people with DSD, including Max Beck, Morgan Holmes, and Kiira Tria, felt wronged by the medical establishment and identified the need to protect the rights of children like them.(Dreger & Herndon, 2009) Specifically, these activists experienced the early childhood interventions on their DSD and the lack of disclosure regarding their condition, to be unjust.<sup>11</sup>

By reinforcing the necessity of truth-telling and full disclosure, these activists have helped challenge the old treatment paradigm under which it was often the case that neither the child (and their future adult) nor the parents were fully educated about the nature of the child’s condition and the purpose and extent of every medical intervention.(Wiesemann, Ude-Koeller, Sinnecker, & Thyen, 2010) When the voices of these advocates are placed in conversation with recent studies that have shown that patients and their parents benefit from disclosure of chronic and stigmatized health conditions,(Blasini et al., 2004; Mellins et al., 2002) their insight has helped support bioethicists call for a re-evaluation of information exchange and the decision-making process in the case of children born with atypical somatic sex development. Current medical trend toward shared decision-making (which will be discussed in Chapter 3) throughout medical practice has also supported this re-evaluation.

In addition to motivating reforms in medical shared decision-making, activists have also helped uncover some biases and assumptions implicit in the care of individuals with

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<sup>11</sup> It’s important to remember that these activists and advocates have never critiqued early interventions medically necessary to ensure physical health. The interventions they describe as harmful are those that are considered cosmetic and elective.

atypical somatic sex development under Money's care guidelines. Asking questions like, "What assumptions about gender are embedded in certain 'classical' approaches to DSD?" or "How much of what medicine recommends is based on medical issues, and how much on value judgments made about extra-medical facts?" encourage a closer examination of the motivations behind specific interventions. For example, it is generally felt that surgery that is performed for cosmetic reasons in the first year of life relieves parental distress and improves attachment between the child and the parents (Baskin, 2004; Crouch, Minto, Laio, Woodhouse, & Creighton, 2004; Farkas, Chertin, & Hadas-Halpren, 2001; Rink & Adams, 1998); the systematic evidence for this belief is lacking (P. A. Lee et al., 2006). There is a similar paucity of information and outcome data on the success of gender assignment and early surgical interventions for individuals with these conditions. Some data that reports on these issues does exist (Brinkmann, Schuetzmann, & Richter-Appelt, 2007; Crawford, Warne, Grover, Southwell, & Hutson, 2009; Creighton, Minto, & Steele, 2001; Crouch et al., 2004; Hurwitz, 2010; Sircili et al., 2010; Wisniewski et al., 2001; Yang et al., 2010), however, findings often contradict each other and use different standards of analysis, making comparison difficult. Hurwitz summarizes the difficulty of interpreting these findings:

"The number of reports is small and the methods of outcomes analysis are varied. Significant problems in outcomes methodology include premature reporting of results, lumping of outcomes of patients with DSDs or ambiguous genitalia, arbitrary and artificial grading systems, lack of standardization of questionnaires, rare documentation of preoperative anatomy, and outcomes based on subjective assessments by surgical and medical providers and by parents. By the time the long-term data are published, procedures and principles of management that were state-of-the-art at the time may have been replaced by newer approaches, thus reducing the importance of those data in contemporary management." (Hurwitz, 2010)

Due to the paucity of valuable and conclusive outcome data, considerable concerns and objections about the use of surgical interventions to “normalize” the ambiguous genitalia of people with DSD have been voiced.(Chase, 1998; Diamond & Sigmundson, 1997) Some have even gone as far as to call for a “moratorium” on surgical interventions until outcome studies are performed with greater precision.(Kipnis & Diamond, 1998) Exposure of these biases and assumptions has called for a re-evaluation of the medical management of children with DSD. In the last few years, for example, “clinicians have become much more attuned to issues of quality of life, recognizing that providing good cosmetic outcomes or normative gender outcomes are not the same as providing the highest possible quality of life for patients with DSD.”(Accord Alliance, 2012)

Support for, and development of, the arguments brought fourth in the intersex rights movement can be credited to a number of different players: Activists, scholars in gender studies, law and bioethics, as well as some medical practitioners have all played a role in the push for a new paradigm of care. Additional credit needs to be given to advocacy groups, support groups, and educational forums specific to this patient population. These groups have provided people with DSD and their families with safe spaces to gather information outside of the medical field and triage hardships specific to DSD through collaboration, while also bringing these issues into public social discourse.

Examples of these groups include the Intersex Society of North America (ISNA), Coalition for Intersex Support, Activism, and Education (CISAE), Bodies Like Ours, MAGIC Foundation (for conditions that affect children’s growth, including some DSD), Accord Alliance, DSD Families, Advocates for Informed Choice (AIC) and other diagnosis-specific support groups like Androgen Insensitivity Syndrome Support Group (AISSG), and CARES

Foundation (for individuals with congenital adrenal hyperplasia), Hypospadias and Epispadias Association, Klinefelter Syndrome & Associates, Turner Syndrome Society of the US, MRHK Organization, Inc. (for women with Mayer Rokitansky Kuster Hauser Syndrome, also known as mullerian agenesis, vaginal agenesis, or the congenital absence of a vagina) and XY-Turners (for individuals with mixed gonadal dysgenesis sometimes referred to as XY-Turners, mosaic XY/XO, or simply XY/XO).

We can also attribute changing social acceptance toward DSD to its presence in the media and public sphere. Two books in particular took the public by storm. In 2000, John Colapinto published "As Nature Made Him: The Boy who was Raised as a Girl." The book recounts the life experience of David Reimer who was treated by Money after a botched circumcision destroyed his penis. Believing that children were gender-dymorphic until the age of eighteen months, Money re-assigned infant David as a girl and subjected him to genital surgery to create a vagina where his penis and testicles used to be. Colapinto's work tells the story of Reimer's transformation and ultimate gender reassignment back to a man. Not only did the book directly speak to the medical community and Money's work, but it also became a New York Time's Bestseller and introduced the world of DSD to the general public. With Reimer's story acting as a primer for social attention, Jeffery Eugenides's 2002 fictional book, "Middlesex: A Novel," about he life of a person with DSD became a Pulitzer Prize-winner. These books brought the existence of people with DSD to the attention of the general public and in doing so acted as educational tools for hundreds of thousands of lay people who had never before thought about DSD.

Many changes to DSD care have occurred in the last two decades. According to a working group, known as "Bioethics and Intersex," (which includes members of patient



support groups (persons with DSD/intersex and/or parents); bio-ethicists; specialists in pediatrics and adolescent medicine, surgery, urology, obstetrics and gynecology, endocrinology; a psychologist and psychotherapist; a specialist in medical law; and a medical sociologist) the four main scientific and social factors that led to a thorough reappraisal of former treatment regimes in DSD care were: (1.) Increasing scientific knowledge about sex determination and differentiation and the complicated interaction of genotype and phenotype; (2.) Modern society's decreasing rigidity about the nature of gender identity and gender roles following the LGBT movement (and other identity politic movements) and the development of the intersex rights movement; (3.) A growing unease about paternalism in medicine and support for the right of patients to know information with full disclosure; and (4.) A growing importance of patient support groups in the assessment of medical interventions and the awareness these groups brought to these conditions in the media and public arena.(Wiesemann et al., 2010)

In 2005, the Lawson Wilkins Pediatric Endocrine Society (renamed the *Pediatric Endocrine Society* in 2010) and the European Society for Paediatric Endocrinology convened 50 international experts in the field of "intersex" care for a consensus conference to review the management of these disorders. While developing an outline of suggested principles to guide care, participants took into account progress in diagnosis, surgical techniques, and understanding of psychosocial issues, as well as recognition and acceptance of the place patient advocacy now plays in these discussions. In 2006, their

suggestions<sup>12</sup> were published in the journal *Pediatrics* under the title “Consensus Statement on Management of Intersex Disorders.”

The Consensus states that “optimal clinical management of individuals with DSD should comprise the following: (1) gender assignment must be avoided before expert evaluation in newborns; (2) evaluation and long-term management must be performed at a center with an experienced multidisciplinary team; (3) all individuals should receive a gender assignment; (4) open communication with patients and families is essential, and participation in decision-making is encouraged; and (5) patient and family concerns should be respected and addressed in strict confidence.” The Consensus Statement also proposed a change in terminology from “hermaphrodite,” and “intersex” to “disorders of sex development” (DSD), discussed various aspects of the investigation and management of DSD, examined existing outcome data in DSD studies and their implications for clinical care, and made recommendations for future studies – one of which was further “evaluation of the effectiveness of information management with regard to timing and content.” (Lee et al., 2006)

Many of these recent changes in the medical management and care received by individuals with DSD were spurred by the suggestions posed in the Consensus Statement. The Statement can be thought of as an outline of the new paradigm for DSD care. While a number of institutional barriers resist the field-wide practice of this new paradigm (Dreger & Sandberg, 2010), DSD care has made major progress in the past twenty years. But there is still much work to be done. Controversy regarding the ethics of early, value-based

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<sup>12</sup> The Consensus Statement is not a set of practice guidelines or used to define standard of care. Rather the Consensus Statement defines a list of suggestions that should guide clinical care of individuals with DSD. The American Academy of Pediatrics (AAP) endorses these principles as a “policy statement.”

interventions and whether or not parents have the right to make these kinds of decisions for their children still exist. With the data currently available, there is little to no evidence to determine if a parent's decision will be *right* or *wrong* – regardless of the criteria with which we judge these distinctions. Rather, debate regarding of the parental decision-making process focuses more on if parents are making *good* or *bad* decisions for their child. Are parents getting the information they need to make the *best* choice for their child? To answer this question we need to take a closer look at the ethics of medical decision-making in this context.

- Chapter 2 -

**Taking a Closer Look at Informed Decision-Making**

*I was discharged from the hospital and when I got home I was brave enough to look up her condition online. It was not something I wanted to go to the Internet for, but I needed more information. I think the one thing I've really struggled with is having to pick and chose specific things from different resources to get quality information that is applicable to my child...*

*...So then the urologist was called in and, like a ray of sunshine, said "I can fix this." She told us that she could fix her genitals in one surgery and after surgery everything would be fine. We had to wait a bit for her to gain weight, but at one year old she had her first surgery. At one of her follow-up appointments we found out the scaring didn't heal quite right. So then, at two years old, they had to go in and do a little repair. Right now things look good, but I'm still concerned with what's going to happen in the future. Some days I worry about what are things are gonna look like as she gets older. I wonder, will people treat her differently? Will she feel like a boy? Will she like boys? Things are functioning perfect right now, but the surgery has only fixed so much. I guess we'll just have to cross that bridge when we get there.*

Quality information exchange (or the lack there of) and disclosure of information (or lack there of) are two topics that have had the most influence on the development of the new treatment paradigm. Adult patients have described the lack of disclosure and the lack information they (and their parents) have received from practitioners as harmful. This has caused some to wonder if – given this lack of information – parents *can* make informed decisions about their child's care. Others have even wondered if – given the lack of evidence-based outcome data, in general, and the child's right to autonomous choice – parents have a *right* to make elective decisions about their child's care. The answers to these questions are central to a discussion of informed consent. This chapter will take a closer look at the informed decision-making process and informed decision-making specific to DSD.

## FOUNDATIONS OF INFORMED CONSENT

Informed consent is an ethical and legal concept grounded in the fact that competent patients have the right to make decisions about their medical treatment, including the right to deny medical treatment. Put into practice, informed consent is a *process* where the healthcare provider supplies information to *inform* the patient and the patient then *consents* to a given intervention after evaluating the provided information.(Devettere, 2010) Informed consent includes education, discussion and analysis by all parties throughout the process – all of which must occur in the absence of coercion. The signing of the informed consent document is a legal acknowledgment that the process did in fact occur, but it is not the informed consent itself.(Devettere, 2010) What exactly this process entails will be discussed in the next section. First, it is important to understand the moral and legal foundations of the informed consent process.

It is commonly held that the three guiding principles for informed consent are respect for persons, beneficence, and justice.(Devettere, 2010; Faden & Beauchamp, 1986; National Institute of Health, 1979) While this tri-part distinction has its value in some discussions, the division is somewhat misleading with regard to informed consent in medical decision-making. In this context, discussions about justice generally appear when it is believed that someone's legal or moral rights have been violated in the process of informed consent. For example, if a person did not receive information that they deserved to know (i.e. that was fair, due, and owed) in order to make an informed decision, then they were not treated justly. The moral duty to provide individuals with all the information they need to make an informed decision is grounded in a moral duty to respect that person's right to autonomy – the right to make uncoerced decisions for him- or herself. So as it turns

out, the moral principle that demands just action in medical decision-making is actually grounded in a respect for persons.(Faden & Beauchamp, 1986) For this reason, respect for persons and beneficence present the majority of the moral and conceptual problems inherent in informed consent.<sup>13</sup>

Respect for persons entails “treating people as autonomous agents capable of deliberation of personal goals and of acting under the direction of such deliberation.”(National Institute of Health, 1979) It is important to make a distinction here between an autonomous *person* and an autonomous *choice*, for informed consent in medical decision-making is concerned with the latter. Autonomous persons – free persons who have the capacity to be independent and who are in control and responsible for their own decisions and actions – can make non-autonomous choices due to ignorance and coercion. Unintentional (ignorance) or intentional (coercion) underdisclosure on the part of the information provider or the provider’s failure to recognize a patient’s refusal of medical interventions are failures to *respect* the patient’s (or in the case of DSD, the parents’) right to autonomous choice. To respect a patient’s or parents’ autonomous choice is to “recognize with due appreciation that person’s capacities and perspective, including his or her right to hold certain views, to make certain choices, and to take certain actions based on personal values and beliefs” after being provided with all necessary information.(Faden & Beauchamp, 1986) The demands that respect for persons place on healthcare providers are not as straightforward as one might initially think. As we will see, it is not always clear what information must be delivered by the practitioner to respect

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<sup>13</sup> Additionally, these problems are not justice-based nor do they directly confront issues of social justice and for that reason the principle of justice tends to take the back seat in these discussions.

their patient, and/or respect for persons will sometimes be at odds with the other guiding principles of informed consent – namely, the principle of beneficence.

The medical principle of beneficence states that the practitioner's goal in healthcare is to promote the welfare of the patient.(Devettere, 2010) This principle is grounded in the practice's foundational maxim *primum non nocere*: "Above all, do no harm."(Faden & Beauchamp, 1986) The principle also demands that practitioners' actions "maximize benefits and minimize possible harms."(National Institute of Health, 1979) The principle of beneficence does not always work in concert with the principle of respect for persons. For a physician may be caught between her desire do what she believes is medically best for the patient (beneficence) and acknowledging the patient's own preferences about treatment and the decision-making process by presenting alternative options that may have worse outcomes (respect for persons). Under Money's optimum gender policy, beneficence was sometimes placed above respect for persons as his policy advocated that physicians withhold the information about the child's condition that could lead to parental uncertainty about the child's gender assignment. Money wanted patients to have the best outcomes and according to his treatment policy, it was sometimes necessary to withhold information.

It has been argued that this portrayal of the conflict that can exist between respect for persons and beneficence is too simplistic in its conceptualization of the latter.(Parascandola, Hawkins, & Danis, 2002) Those who argue this point claim that the definition of beneficence is to promote the *welfare* of the patient. In the argument outlined above, "welfare" is confined to what is "medically best," i.e. the choice or action that leads to the best health outcome. This is a very constricted conceptualization of well-being. What

really promotes overall well-being may involve allowing patients to make decisions even at the *expense* of what is medically best. Studies have shown that involving patients in the decision-making process has extremely beneficial outcomes such as enhanced patient satisfaction, adherence to treatment plans, greater confidence in health care recommendations, symptom resolution, psychological well-being, and overall quality of life. (S. Ford, Schofield, & Hope, 2003; Fraenkel & Peters, 2009; Greenfield, Kaplan, Ware, Yano, & Frank, 1988; Joosten et al., 2008; Kaplan, Greenfield, & Ware, 1989; Loh et al., 2007; Légaré et al., 2010; Ward et al., 2003) These findings support many reasonable theories of well-being that argue that self-determination is in itself an important aspect of well-being. (Parascandola, Hawkins, & Danis, 2002)

Today it is generally accepted that when beneficence and respect for persons seem to be at odds with each other, the latter should be used to guide practice. But this debate is far from over. Stripped down to the core of the debate, what is really being asked is “how much information is the best amount of information to provide parents?” While it is obvious that providing too little information can have detrimental effects on decision-making, recent studies have shown that too *much* information can also have detrimental effects. (Edwards & Yahne, 1987; Schwartz, 2011; Ubel, 2002) Scholars do not agree how much information is the right amount of information to provide patients in order to honor the principles of beneficence and respect for persons. Deciding how little information is too little and how much information is too much is not as easy a task. Until we better understand how and why information quantity affects people’s decision-making, the burden of proof is again placed on those who argue for withholding information. (Parascandola et al., 2002)



One could argue that the requirement that physicians provide patients with information stem from the higher-order moral requirement than respect for persons and beneficence. That is, to treat people as beings with intrinsic value. One of my favorite philosophers, Immanuel Kant, claims that we should “act so as to treat people always as ends in themselves, never as mere means.”(Kant, 1998) What does Kant mean when he says this? He is calling attention to the ability people have to govern themselves by using their reason to set their own goals. We value people as the kinds of beings that can do this when we treat them (and more specifically, what they can rationally endorse) as constraints on our own goals -- limits on what we can and cannot do to achieve our own ends. Put another way, we treat people as ends in themselves when we treat them as beings with their own purposes, and not merely as useful tools we can employ to satisfy *our* personal purposes. We use each other as means everyday: Patients can use physicians as a means to improve their health, and conversely, physicians can use patients as a means to make a living, so as not to treat each other as *mere* means, constraints are placed on how you use people. In particular, to treat someone as an end in the context of medical decision-making is to treat her as a rational being capable of making informed decisions regarding her medical care. To make informed decisions, she needs information. What information do healthcare providers need to provide to a patient in order to respect her as an end in herself? Let’s find out.

#### REASONABLE PEOPLE AND REASONABLE PARENTS

The requirements of information exchange in informed consent are set by “the reasonable person standard.” The reasonable person standard states that a physician must

disclose all information that a “reasonable person” would consider relevant to the patient’s decision. This is generally thought to include information regarding (1.) the nature of the condition and its prognosis, (2.) the nature and purpose of the proposed intervention, (3.) *common* risks of the proposed intervention, (4.) *remote* risks with serious consequences, (5.) the likelihood of success of a given intervention, including short- and long-term outcomes, and finally (6.) the risks, benefits and uncertainty associated with alternative interventions and non-interventions.(Karkazis, Tamar-Mattis, & Kon, 2010)

Of course no two patients are the same in all respects. It is reasonable to expect that different patients would desire different information or information presented in different ways based upon their personal situation. As one physician put it, “I do not care for “average patients”; I care for individual patients. [...] The physician’s duty is to provide the best available current information to individual patients and to assist them in making their own best decisions.”(Karlavish, Fox, & Pearlman, 2002) Recent focus on patient-centered care in the US, which places individual patients (not standardized “objective” cases) at the center of decision-making, has introduced the expectation that physicians provide information that fulfills the “subjective person” standard as well. This means that physicians must provide all information needed by a *reasonable person* – the “objective person” – but in addition they must also provide any and all information that the particular person – the “*subjective*” person – might find pertinent to decision-making.(Faden & Beauchamp, 1986) The subjective person standard implies that a health care provider needs to inform a patient in terms of what the patient *actually* wants to know, not just what a reasonable person *should* want to know.(Holmes-Rovner & Wills, 2002) This process is more individualized, and acknowledges that every patient is different; every person comes

with his or her own set of values and concerns and his or her own biases and assumptions – all of which are influenced by larger socioeconomic factors, power dynamics, language barriers, gender differences, and cultural context. Only if healthcare providers are sensitive to these differences can they provide patient-centered care.

In the case of early DSD interventions, however, it is not the patient (the infant or young child) who is participating in the process of informed consent. Rather, parents are responsible for making these early decisions for their children. This allocation of decision-making power is commonly referred to as “proxy-consent” and it occurs when the patient is unable to make rational decisions on his or her own behalf. Children (especially young children – less than five years of age) lack this capacity to participate in decision-making, and therefore require their parents to act as proxy decision-makers.

In 1995 the American Academy of Pediatrics (AAP) Committee on Bioethics proposed that the term “proxy-consent” actually has a limited direct application in *pediatrics*: The Committee points out that the word “consent” literally means “to feel or sense with;” thus, consent implies the decision is made to address the feelings of the decision-making, rather than to address the patient’s needs and desires. *Proxy-consent*, then, “poses serious problems for pediatric health care providers who have a legal and ethical duty to the child to provide competent care based upon what the *child* needs, not what someone else expresses.” In an attempt to put the focus back on the needs of the child, and away from the needs/desires of the parents, the Committee of Bioethics proposed the term “informed permission” as a replacement for “proxy-consent.” (American Academy of Pediatrics: Committee on Bioethics, 1995) This nomenclature revision acts to place the child at the center of decision-making where decisions are made based upon the *child’s* best

interests. This is an important point to remember when it comes to the medical care of all children.

More valuable to the care of children with DSD, in specific, may be a different nomenclature revision. That is, changing “informed consent/permission” to “informed choice” or “informed decision.” Both of the terms “informed consent” and “informed permission” imply a passive role on the part of the patient or parent where they *allow* something to happen: the physician *informs* (active) the patient and the patient *consents/grants permission* (passive) to an option presented by the provider’s information. With a recent focus on shared decision-making where providers and patients *share* the responsibility of deciding on the best course of action, the terms “informed choice” or “informed decision” have been suggested to replace “informed consent.” (Charles, Gafni, & Whelan, 1997) This nomenclature implies an active role for both providers and patients where practitioners *inform* (active) patients or parents about treatment **alternatives** and patients or parents *decide* (active) (in collaboration with their healthcare providers) on the best course of action. A central ethical principle behind informed patient choice is that the information is being given in order to *enhance* choice. (S. Ford et al., 2003) An overarching point that needs to be made throughout the discourse of DSD healthcare is that, in many situations, non-intervention is an appropriate alternative to medical treatment or intervention. A decision is defined as a choice in a course of action among a set of options with the intent of achieving a goal. (Baron, 2000) The rhetoric of “informed decision” therefore implies the availability of multiple options from which the patient or parent decides. To emphasize the points that parents have options and that they are active

members in the decision-making process, I will use the term “informed decision” throughout the rest of this paper instead of “informed consent.”

Since multiple treatment options are appropriate in certain value-based (rather than consensus-based) decisions that parents must make regarding the care of their child with DSD, it is not always clear what decisions are in the child’s best interests. Some people have wondered whether parents have the right to make decisions regarding interventions for their child when consensus is lacking as to which decisions are in the child’s best interests. This argument is outlined by Wiesemann et. al. as follows:

“Parents’ surrogacy should extend merely to decisions that are in the objective best interest of the child.(Diamond & Sigmundson, 1997; K.-K. Ford, 2001) The appearance of the external genitalia in DSD generally causes no medical problems or immediate health threats to the child. Moreover, most surgical interventions are irreversible and may restrict later options for sex reversal. As it is difficult to find out what is in the best interest of the child, nobody, the argument goes, should decide on these important questions except the patient him/herself. This position entails postponing minor interventions at least until the age of 5 or 6, when the child is able to express preferences and major interventions until the age of 12 to 14.”(Wiesemann, Ude-Koeller, Sinnecker, & Thyen, 2010)

At first glance this argument against early interventions seems to promote respect for the child’s bodily integrity and autonomy in a way that parental permission would violate. For non-intervention on the child’s body upholds this cultural idea that only a body free of intervention is a body with integrity(Slatman, 2011; Tjaden, Tong, Henning, Groothoff, & Craig, 2012) and leaving the decision up to the child’s adult self allows the individual to be the autonomous agent making decisions for him- or herself. But upon further review, we see that respect for these principles is more complicated than this. Wisemann et. al. developed the following counter-argument:

“According to the maxim of postponed informed consent, only the adult would be truly able to determine the best interests of the child he or she was in a former time. But this child no longer exists, and its interests, thus, can no longer be respected. For example, a child with incomplete Androgen Insensitivity Syndrome<sup>14</sup> raised as a girl might in puberty face the effects of virilizing hormones. Hormone suppression has to start before puberty to avoid the effects of male-typical pubertal development and to prevent major disturbances in gender identity. Postponing this decision to the age of consent, however, means closing an important window of opportunity for the child. The future adult’s consent, thus, will be meaningless, because no decision will undo the consequences of a waiver of treatment in the past.” (Wiesemann et al., 2010)

One may argue that this is an example where the child’s best interests are clear: hormone suppression should be induced. The point of this example is that when it comes to DSD, decisions can be time sensitive. A parent in the University of Michigan’s research study explained her thought process about this situation in a slightly more nuanced way:

*For a while I did contemplate not having surgery at all until she was old enough to make those decisions for herself. Probably the biggest reason we were uncomfortable with that was because we were told that she needed to make that decision by the time she turned 8 or 9 if she was going to be raised female and avoid any masculinization. We weren’t convinced that a child at the age of 8 or 9 could really make that decision.*

Important windows for intervention may be lost if the decision is postponed for the future adult. Children do have a right to bodily integrity and respect for autonomy, but autonomy is grounded in the ability to consent and those individuals not capable of autonomous choice (including children) are unable to make decisions. When people (children) are unable to make these kinds of decisions, a proxy decision-maker (such as a parent) is designated to make the decisions for them.(Chappuy et al., 2006; Devettere,

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<sup>14</sup> As stated by the AIS Support Group: “Androgen Insensitivity Syndrome (AIS) occurs when an individual has one X and one Y chromosome, but the body is unable to respond to androgens (male sex hormones). During prenatal development, an individual with complete AIS (CAIS) will develop female external genitalia, while a person with partial AIS (PAIS) may have external genitalia that appear mainly female, or mainly male, or anything in between.” (AIS-DSD Support Group: For Women and Families, 2012)

2010; Ross, 2004) Parental permission shows respect for the child to the extent that it serves to protect the child's best interests.(Ross, 2004) If it's true that parents can serve their child's best interests by making early decisions for them, then Ackerman is correct that "we fool ourselves if we argue that we have fulfilled our moral duty by standing aside and asking the child to decide."(Ackerman, 1979)

Both ethical and legal discourse grant that parents have the right to make these decisions for their child for a number of reasons. It is widely assumed that parents are best able to make decisions that promote the best interests of their child because they (1) "care deeply about the welfare of their children and know them and their needs better than others do"(Chappuy et al., 2006), (2) "take first-line responsibility in defining what might be best for the child, and this may rightly vary according to their individual experience and life style, cultural expectations, and beliefs"(Wiesemann et al., 2010), and (3) parents "bear the consequences of treatment choices for their dependent children."(Chappuy et al., 2006) Although these reasons focus on parental duty to serve the child's best interests, some modern philosophers argue that parents are justified to make these decisions for their own ends(L.F. Ross, 1998): within limits, parents have a right to "raise their children according to their own standards and values and to seek to transmit those standards and values to their children." Parents also have a right to "promote family intimacy," which requires significant freedom – including the freedom to make important decisions about the welfare of its dependent members.(Buchanan & Brock, 1989) The process of informed decision-making, then, "not only promotes the best interests of the child, but respects legitimate parental interests of making decisions for their child and family."(Chappuy et al., 2006)

From this point on, then, I will assume that parents have a right to make decisions for their child, including those decisions that are not of consensus value.

### CHALLENGES TO MAKING INFORMED DECISIONS

Parents have a duty and a right to make informed decisions regarding the medical care of their children. Becoming informed about these decisions is a process that requires providers to deliver information to parents and parents to comprehend this information. In order for a parent to make an informed decision, three qualities must be fulfilled: Parents need to be in possession of all the relevant facts at the time of decision-making, they need to possess adequate reasoning faculties, and they must make the decision free from coercion. Achieving these three qualities can be a challenge to both providers and parents. For the sake of discussion, let's just assume that these decisions are being made free from coercion, and focus on the first two qualities. For even with just these two, things get tricky: Providers must appropriately deliver information even when it is difficult to do so, and parents must possess the capacity to comprehend and analyze this information at times when there are particularly unfit to do so. In this section I'd like to call attention to three of these challenges. Namely, provider's ability to deliver information in a way that fulfills the subjective person standard, provider's ability to deliver information in the face of uncertainty, and parents ability to reason during times of stress and anxiety. Each of these concerns needs to be taken seriously to ensure that informed decisions are made.

To begin, the subjective person standard requires that physicians provide all the information that a *particular* parent may find pertinent to decision-making, given their specific beliefs, values, and situation. (Faden & Beauchamp, 1986) But the information that



parents need and want provided to them by the healthcare team is not always clear.(Tamar-Mattis, 2009) As was highlighted when discussing the subjective person standard, it is important to remember that all of these discussions are happening in a larger context where people of different cultures bring with them culturally specific values and concerns and a native language that may not be the same as the healthcare providers'. Additionally, parents' desire for information type and quantity is not always the same. For what one parent deems relevant to his or her decision-making process may be of no importance to another parent. Healthcare providers are responsible for discerning these differences between parents. Yet, research evidence suggests that when physicians infer patient preferences for information and for participation in treatment decision-making, they often fail to get them right(C. Charles, Gafni, & Whelan, 1997; Ryan, 1992; Strull, Lo, & Charles, 1984; Waitzkin, 1991) These challenges reinforce the fact that physicians must pay careful attention to parents' individual informational needs so that they can provide parents with the individualized information they need to make informed decisions.

Studies have shown that in clinical situations with substantial uncertainty, physicians find it even more difficult to uphold the standards of information exchanged necessary to parents making informed decisions. In particular, this study found that healthcare providers are more likely to withhold potentially valuable information to patients when interventions are considered to have one or more of the following qualities: they are severe, common, elective or non-therapeutic, experimental, controversial, and/or when intervention outcomes are uncertain. (Parascandola et al., 2002; Tamar-Mattis,

2009)<sup>15</sup> Uncertainty manifests itself in a number of ways: It can stem from a lack of scientific data on a given condition's etiology, the interventions and their outcomes. It can also occur in the presence of known data – as there is always a given degree of ambiguity in applying known data to a particular case. Since “uncertain knowledge is usually complex and difficult to communicate, many physicians revert to nondisclosure and nondiscussion or oversimplification when discussing the situation with patients.”(Parascandola et al., 2002) Physicians need to be aware of these tendencies and provide parents with this information even if it is difficult to do so.

Another reason why physicians may find the delivery of this kind of information difficult is that they fear that they may harm their patients by providing them with uncertain information. Physicians worry that their patients “may be greatly upset or overwhelmed by discovering the uncertainty of their prognosis or treatment success.”(Parascandola et al., 2002) Uncertainty is a source of substantial stress and anxiety for many patients – especially for patients with serious long-term illnesses such as cancer, heart disease, AIDS, and diabetes, who may endure feelings of vulnerability and ambiguity about future years on end.(Parascandola et al., 2002) Parents of children with DSD have also indicated that they too experience high levels of shock, stress and anxiety after their child is diagnosed with a DSD.(Crissman et al., 2011; Duguid et al., 2007; Fedele et al., 2010; Jürgensen, Hampel, Hiort, & Thyen, 2006; Lev, 2006; Malmqvist & Zeiler, 2010; Sanders, Carter, & Goodacre, 2011) This parent with DSD said:

*The most stressful thing was just that her condition was rare and there was a lack of information. I think finally one of the doctors is printing some stuff out*

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<sup>15</sup> Severe” (i.e. irreversible), “uncertain,” “elective,” “controversial.” These are all words we hear repeatedly in the DSD intervention rhetoric. As discussed in the last chapter, DSD management has a history of non-disclosure.

*for me on the computer, you know, that I can read. But the most stressful thing is just not being presented with clear-cut information.*

Some parents even express *clinical* levels of stress in response to their child's DSD.(Duguid et al., 2007) Parents state that this stress is, in part, due to the paucity of information about their child's conditions and the next steps they should take, the fear that their child will experience stigma in the future because of their condition, and an inability to talk to others about their child's DSD.(Crissman et al., 2011)

In Eder et. al.'s study of parents of children with Leukemia he found that parent's most frequently cited suggestion for improving informed consent was to give them more time to make decisions about participation in clinical trials. Parents who offered this suggestion stated that they needed the opportunity to cope with their emotions before making the decisions about their child's care. (Eder, Yamokoski, Wittmann, & Kodish, 2007; C. Simon et al., 2001) When parents have relatively little time to make decisions regarding their child's medical care because the decisions are time-sensitive, physicians may push parents to make decisions faster – as was described in Eder et. al.'s study. In the case of DSD management, however, some decisions are not time-sensitive and *parents* are the ones eager to make decisions. One reason parents have used to explain this feeling of urgency is that they must talk to others about their child and they have a hard time finding the right words when their child does not have a gender assignment:

*After I delivered they said, "Your son has a severe hypospadias and an undescended testicle." I guess that was the red flag that we need to check to make sure that he was an unknown gender then. So he was whisked away to ultrasound, whisked to x-ray, and where ever else, and my husband's still not back! I was trying to call him, and I'm like "Don't tell anybody we had a baby yet, because I don't know what he is. They can't tell me if it's a boy or a girl." That was just the worst.*

As one parent described in another study, “it was better to make some decision and revise it later than leave the child's gender open-ended any longer.”(Jürgensen et al., 2006)

Time is relative. What may seem like unlimited amount of time to outsiders because of the nature of some of these DSD interventions, may seem like an emergency to parents.(Monteverde, 2009) This is potentially problematic for physicians as they need to confirm that parents have the *capacity* to make informed decisions (with life-long consequences for the child) while stressed, but also they do not want to alienate parents by delaying their decisions which may cause them to go to a different provider. The 2006 Consensus Statement on Management of Intersex suggests that emotional and psychological support needs to be provided to parents during the decision-making process.(Lee, Houk, Ahmed, & Hughes, 2006) This kind of support in a multidisciplinary team may provide parents with the tools they need to slow down their decision-making process by addressing their feelings of urgency and stress with the help of a psychologist specialized in these issues.

These mental health professionals could be a pivotal part of assessing a parent's decision-making capacity. The definition of decision-making capacity varies somewhat across jurisdictions, but generally embodies the patient's ability to communicate a choice, to understand the relevant information, to appreciate the medical consequences of the situation and to reason about treatment, alternative treatment, and non-treatment.(Appelbaum, 2007; Berg, Appelbaum, & Grisso, 1996; Grisso & Appelbaum, 1998)<sup>16</sup> If a patient (or parent) has the ability to understand, evaluate, and reason about

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<sup>16</sup> In 2007 Appelbaum concluded there are no formal practice guidelines to assess a patient's capacity to consent to treatment from professional societies.(Appelbaum, 2007) As far as I'm aware a standardized set of practice guidelines is still non-existent.

decision-making they are considered to have decision-making capacity.(Devettere, 2010)

This ability to reason is dependent on both cognitive and emotional competence.(Breitsameter, 2010)

Impairments to reasoning and judgment that may affect a person's decision-making capacity include basic intellectual or emotional immaturity, dementia or deliria, incapacitating illness, and/or high levels of stress. Physicians must be aware that their patients and/or parents may have impaired decision-making capacities, and they must be skilled at evaluating that possibility.(Appelbaum, 2007) Sometimes a patient's decision-making *incapacity* is obvious – i.e. the patient is too sick to make a decision, is delirious, or has a permanent handicap – but other times it is much less so. In some cases, for example, impairments like stress may be severely debilitating to a person's decision-making capacity but difficult to identify.

Chappuy et. al.'s investigation into parental consent in pediatric clinical research found that parents demonstrated better understanding when informed consent was sought some time after the child's diagnosis was made, rather than at the same time. Given time to deal with the stress of the situation, parents were better able to understand the aims, potential risks and potential benefits, and the possibility of alternative treatments for a given intervention.(Chappuy et al., 2006) This study shows that a person's decision-making capacity changes with time. This capacity is something that can come and go, or be present in some decisions but not others based upon one's current cognitive and emotional state. Parents may be too shocked and distraught immediately after the birth of their child with DSD to understand the information necessary to make an informed decision about gender

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assignment. Parents have expressed this by saying they experienced and “information overload.” During these times, parents would not be considered to have decision-making capacity because they were lacking understanding. But as Chappy et. al.’s study suggests, this does not mean that they will *always* lack this capacity.

Common sense would suggest that there should be degrees of capacity, for a person’s capacity for decision-making could diminish for a number of reasons and yet not be completely lost.(Devettere, 2010) Parents could be stressed, but not so stressed that they are rendered incompetent to make decisions. Similarly, a child’s capacity for decision-making could be partially developed, but not completely developed. While this may be true, legal considerations dictate that decision-making capacity must be thought of as a threshold: you either have it or you don’t. Unless we “draw a sharp line between capacity and incapacity, there is no way to determine who has the ultimate responsibility to make the final decision about a patient’s care or intervention – the patient or their proxy.”(Devettere, 2010) In order to make informed decisions, patients must have the adequate decision-making capacity. Physicians are responsible for evaluating this capacity and determining if it meets the threshold of necessary cognitive and emotional competence.

No one has ever said that the process of informed decision-making is an easy one – especially when it is complicated by the fact that decisions are being made by proxy-decision-makers with individualized information desires, in the face of clinical uncertainty, and under high levels of stress. When it comes to DSD, there is a lot at stake in these discussions for all parties involved: parents must make life-changing decisions for their child, children must live with these decisions, and physicians must accurately deliver

information and evaluate parents' decision-making capacity else they face ethical and legal liability.

#### DSD-SPECIFIC CHALLENGES TO INFORMED DECISION-MAKING

The challenges of informed decision-making outlined above are not specific to the medical management of DSD – though all of these challenges are potentially present in this context. Given the history of non-disclosure, the controversy, the paucity of evidence-based information, and parents' constant call for more information, it should not be a surprise that informed decision-making for parents of children with DSD has its own hardships. Parents and physicians need to be aware of these challenges particular to DSD informed decision-making so that they can ensure the best decisions are being made for the child. The challenges are as follows: (1.) Excessive provider optimism and failure to tell parents about doctor's level of experience with DSD, (2.) Physicians' failure to tell parents about the controversy surrounding DSD intervention options and present all intervention options, including non-intervention, and (3.) Physicians' difficulty discussing sex, gender, and sexuality. (Tamar-Mattis, 2009) I will expand on them here.

(1.) *Excessive provider optimism, and failure to tell parents about doctor's level of experience with DSD.* Available evidence suggests that in clinical circumstances lacking community consensus, such as interventions for children with DSD, clinicians exhibit excessive confidence in their own recommendations, thereby preventing full consideration of alternatives. (Baumann, Deber, & Thompson, 1991) This phenomenon exists in DSD care. Take this physician's words as an example. As there is little to no outcome data that

upholds his or her statement, this physician expresses an overly positive sentiment toward their intervention technique:

*“Our approach to the clitoroplasty leaves the patient with intact clitoral sensation, painless sexual arousal, a viable and sensate glans clitoris and appropriate erectile function during sexual arousal.”(Tamar-Mattis, 2009)*

Physicians’ excessive optimism can misinform parents about the reality of specific interventions. This parent, for example, interpreted a physician’s words to mean that early surgery will be the fix-all for every aspect of the child’s DSD:

*If we waited until he was 12 to do it, there could be social effects. If he didn’t have the surgery, he would never be able to stand up and go to the bathroom like a normal male would. [Physician] said if we have the surgery, all of those things would be normal for him for the rest of his life.*

A study analyzing the parental experience of making these early decisions found that parents continue to worry about the social effects of their child’s DSD even after surgery(s) have concluded.(Crissman et al., 2011)

The Consensus Statement states that “only surgeons with expertise in the care of children and specific training in the surgery of DSD should perform these procedures.”(Lee et al., 2006) Physicians need to be familiar with the realistic outcomes parents can expect from these interventions, and explain these realities to parents. Only those physicians specialized in this care are fit to do so. For this reason, the Consensus Statement calls for all children with DSD to be cared for by a multidisciplinary team of practitioners specialized in this care.

(2.) *Physicians’ failure to tell parents about the controversy surrounding DSD intervention options and present all intervention options, including non-intervention.*

Some parents take it upon themselves to gather additional information about their child’s DSD in addition to the information that physicians provide them.(Crissman et al., 2011)



This can give them exposure to the larger context of DSD interventions, alternative options, and the controversy regarding cosmetic early interventions. Other parents, however, rely completely on the information given to them at the hospital:

*I got a lot of paperwork from the hospital so I had read that a lot. That explained a lot. I didn't search on the Internet.*

These parents are the ones that could easily fall prey to a telescoped understanding of possible DSD interventions. This telescoped understanding is based on framing, namely a particular schema of interpretation that people use to know and navigate the world based upon a specific set of “filters” to which they are exposed.

According to an influential theory of epistemology known as “standpoint epistemology,” we come to understand the world from a given perspective based upon our culture, gender, socioeconomic standing, age, profession, etc. All the characteristics that situate us in the world shape the lens through which we can know the world, thus giving us access to certain perspectives and simultaneously excluding the existence of others from our knowable universe.

In using a specific discourse – like Western biomedicine – to understand a situation and a specific rhetoric – like diagnostic terms – to describe it, we pigeonhole our understanding of a situation into a given standpoint. Some activists and scholars have pointed out this effect specific to DSD. The 2006 Consensus Statement suggested a change in the nomenclature from “intersex” to “DSD.” There was ultimately a great deal of controversy surrounding this change, due to the fact that the conditions became medicalized via the terminology used to describe them. We live in a culture where diseases and/or *disorders* need a diagnosis and diagnosis implies treatment. (Ablon, 1981; Davis-Floyd, 2003) In the case of DSD interventions, “presumptions, judgments, values and

presuppositions brought by the physician to the identification, diagnosis, and curative procedures create a network of constraints that exclude alternative possibilities. The result is a situation wherein parents, physicians, and intersexed patients have “no choice” but to accept the medical treatment guidelines” (Hester, 2004) – which in the medical field, generally include surgical interventions (especially if a surgeon is the one delivering the information to parents). As one urologist recalls:

*We have not yet had one family who, in spite of multiple discussions and given options, has not wanted to have corrective surgery earlier on rather than wait until post puberty.*

Standpoint anyone? If all of those discussions were founded on biases toward surgical interventions, it wouldn't necessarily matter how many of these discussions parents had.

This is something acknowledged by another two physicians:

**Interviewer:** *Is there the possibility that part of what we perceive as the parent's urgency to have early surgery, could that partially be signaled by our communications with them?*

**Physician 1:** *Absolutely.*

**Physician 2:** *Yeah.*

Some parents of children with DSD explain that doing surgery or not doing surgery was never an option. (Crissman et al., 2011) This could reflect the general tendency for intervention after diagnosis that exists in the medical field. This parent says that she didn't even discuss alternative options other than surgery:

*Right away we said we wanted him normal, you know, as normal as possible, and [surgery] was never even a choice for us. We never discussed it, you know, not doing it.*

Knowing what we do about framing and standpoint epistemology and the fact that all of us bring certain biases with us to conversations, one could make a case that in order to better provide parents with information about *all* available options, surgeons should not

be the ones responsible for speaking to parents about interventions. This is not always feasible in institutions that do not have multidisciplinary teams that specialize in this care. Multidisciplinary centers have psychological support and a better knowledge of larger DSD discourse outside of the medical field, and therefore are better equipped to make sure parents are given all the available information necessary for informed decision-making. In addition, the consensus statement supports the inclusion of advocacy groups in these discussions. These groups could provide another standpoint of information, and only those physicians specialized in DSD care would know of their existence and easily be able to provide parents with information about the groups.

(3.) *Physicians' difficulty discussing sex, gender, and sexuality.* There is stigma attached to discussions about sex, gender (including gender role and gender identity), and sexuality. These are discussions that have historically belonged to the “private” domain, and often when they are transferred into “public” realm people become uncomfortable. This parent expresses the discomfort she feels when talking about her child’s genitals to others:

*It's a little bit of a personal area of the body so it's, a little uncomfortable to talk about. I don't know. I don't want to run around with a banner saying that my child has an issue with that part of his body.*

For a few reasons, healthcare providers need to be comfortable and well versed in discussing these topics with parents. First, providers need to accurately explain how sex, gender, and sexuality relate to the child with DSD. Second, if healthcare providers can convey comfort with the topic, then parents may pick up on that and similarly feel more comfortable talking about their child’s conditions with others. Studies show that both

parents and affected children benefit from this disclosure of the condition to others.(Blasini et al., 2004; Lee et al., 2006; Mellins et al., 2002)

Once again, in order to achieve the ethical standards of informed decision-making, children with DSD need to be cared for by healthcare providers that understand the complexity of this care. Providers need to know what sex, gender, and sexuality are, what they imply about clinical interventions for children with DSD, and how normative understandings of their associations can influence parents' understanding. DSD-specific multidisciplinary teams are best suited to discuss these issues with parents as they are – theoretically – more educated than your average “hard science” doctor about the “soft science” social issues of sex, gender, and sexuality; they have these discussions more often than other practitioners and therefore should be more practiced in talking about these issues; and finally, they are – theoretically – conscious of the complexity that is inherent in these discussions which force a direct interplay between medical care and larger social context.

#### TWO POINTS TO TAKE AWAY

If just two points are taken from this chapter, they should be this: First, parents have a right and a duty to make early informed decisions regarding the medical care of their child's DSD, and second, the process of informed decision-making is wrought with challenges in this clinical context that cause information to be lacking. Despite these challenges, parents must make some early decisions regarding their child's care and it is ethically and legally required that they be provided with all information material to decision-making. Practitioners need to be conscious of the common challenges, and specific DSD-related challenges, that occur in the informed decision-making in the clinical care of

individuals with DSD. If practitioners are aware of these challenges, they can actively work to provide the information that is difficult to deliver and ensure the best decisions are being made for the child.

### - Chapter 3 -

#### **What Shared Decision-Making and Decision-Aids have to Offer**

*One day, when I came back to the hospital to visit my child, they told me that there were some decisions that my husband and I would need to make. Some of them, they said, were pretty straight-forward and needed to be done for health reasons. They told me that other decisions would be my choice. In particular, they said there was extra skin on my daughter's genitalia and she would have to have surgery. I could either wait until she got older and have her choose whether or not to do the surgery or I could decide for her now. I don't even know why he gave me the option. It was never even a choice for us. The minute she was born we knew this had to be fixed. We wanted her normal, you know, as normal as possible. We never even discussed not doing the surgery.*

An informed decision is a quality that is achieved through a process of information gathering, exchange, and analysis. There are different ways this process can be approached to achieve this quality in a decision. Today, the standard decision-making approach used in clinical practice is known as “shared decision-making.” Where informed decision-making is a quality achieved through the process of information exchange, shared decision-making is a style of taking that information and using it to come to a final decision. There can be informed decisions that are not shared, and shared decisions that are not informed. Today, good decisions in the medical context are thought to include both this quality and style – that is, they are “informed shared decisions.”

In the last chapter focused on the process of informed decision-making that leads to informed decisions. This chapter will discuss the process of shared decision-making where parents and healthcare providers work together to come to a final decision. This chapter will also discuss how decision-support tools known as “decision-aids” can facilitate informed shared decision-making. Finally, this chapter will investigate what the process of shared decision-making and the use of a decision-aid could offer parents of children with DSD while they make these early decisions for their child.

## CHANGING TIDES AND THE MOVE TOWARD SHARED DECISION-MAKING

As always, we must begin with a little history to set the stage.

Current standard of care advocates the use of evidence-based medicine to guide decisions regarding medical interventions. The rationale for treatment interventions with evidence-based medicine rely on data from population studies rather than biological theory and prediction based off of that theory; it focuses on *whether* evidence shows that a treatment works, rather than relying on biological theories about how a treatment *might* work.(Barratt, 2008) So what does this mean in practice? Here is an example provided by Alexandra Barratt used to describe the value of evidence-based medicine:

A recent example of the benefit of evidence-based medicine has to do with the treatment of acute head injury from trauma such as motor vehicle accidents. For many years doctors were taught that when there is trauma to the head, the brain swells and that can cause long term damage and death. Thus, intravenous steroids are given to reduce the swelling. In the 1990s the UK MRC funded the CRASH placebo trial to test whether this theory was a sound basis for treatment. The trial recruited about 10,000 patients – it was designed to recruit 20,000 around the world but it had to be stopped early. When this trial reported its findings in the Lancet in 2003 it showed an increase in the risk of death (3% higher in absolute terms and 18% higher in relative terms) in the group receiving steroids. Steroid infusion should have reduced death and disability, but the trial evidence showed convincingly that it did not. An accompanying editorial estimated that this ill-founded treatment killed about 2,500 people worldwide each year for the 30 odd years during which it was in use – about 75,000 people in total.(CRASH Trial Collaborators, 2004 in Barratt, 2008)

The use of current, best, evidence-based medicine to guide medical practice is now widely accepted and is the ideal standard of care.(Sackett, Straus, Richardson, Rosenberg, & Haynes, 2000)

I say “ideal” because, in some cases, evidence just doesn’t exist, and thus cannot be used to guide practice. As discussed in Chapter 1, when it comes to DSD, outcome data is lacking and the data that does exist is often contradictory or uses different standards of

evaluation (thus, making comparison of the data difficult).(Hurwitz, 2010) For example, there are few to no control trials that compare early vs. late genital surgery or different techniques.(Lee et al., 2006); the standards used to determine the success of gender validating surgeries have changed through time, e.g. successful peno-vaginal intercourse to health-related quality of life (Dreger, 1998); and the data that we do have does not always extrapolate. For example, we have some information about the frequency and outcome of gender assignment of certain conditions, but because DSD encompasses so many different phenotypic and physiologic aspects what is true for one condition is not necessarily true for another. In the face of little data, lots of controversy, stress and uncertainty, parents must make early decisions for their child with DSD. Both parents and physicians have the benevolent goal of trying to determine what is best for the child and take measures that bring about that goal, but what is “best” is not always clear, and what decisions will lead to that outcome are not necessarily straight forward. Thus, parents must make decisions based upon their personal values and beliefs, with little evidence-based outcome data to guide their decision-making process.

As briefly mentioned Chapter 2 when discussing the standards of the *subjective* person, this kind of decision-making that acknowledges the value of subjective criteria is known as a patient-centered approach. This model of care places a strong emphasis on individual patient participation in clinical decision-making by taking into account each patient’s needs and preferences.(S. Ford et al., 2003) This bio-psycho-social approach combines known evidence-based medicine *and* a patient’s personal and contextual elements to determine the best course of action for that specific individual, in that specific decision. This approach acknowledges that each individual person could reasonably make



different decisions. Ultimately, the practice used today, known as *shared decision-making*, grew out of this effort to involve parents in decision-making as part of the larger ideological trend toward patient-centered care.

Reflected in the quip “Doctor knows best,” the past model of decision-making left little room for patients’ input in making decisions regarding their health care. This model was known as the “paternalistic model” of medical decision-making. Under this model, it was argued that because the physician knows more than the patient about medical care, interventions and their outcomes, then the physician must know what is best for the patient – better than the patient could know for themselves. It was thought that if the physician truly cared for the patient, then he would simply do what was in their best interests, medically. “If this meant doing things without the patient’s knowledge and consent, so be it.” (Devettere, 2010) This model places patients in the passive, dependent role vis-à-vis the physician as an expert with authoritative knowledge and skill.

The paternalistic model of medical care has its roots deep in the history of medicine. Some claim that its ideals stem from the original version of the Hippocratic Oath, written by Hippocrates in the late 5<sup>th</sup> century BC. The oath has physicians pledge:

*I will apply dietetic measures for the benefit of the sick according to **my** ability and judgment; I will keep them from harm and injustice.*

By omitting statements that imply value in *patient’s* judgments, the oath conveys a sense of medical paternalism where the physician’s (and only the physician’s) ability and knowledge are the ones that matter.

In the most extreme cases of medical paternalism, a physician will simply tell the patient which intervention to use. In the less extreme cases, the physician will give the patient limited, potentially biased and/or deceiving information and will encourage the

patient to consent to what the physician considers best.(Emanuel & Emanuel, 1992)

Money's optimum gender policy sometimes utilized the latter form of medical paternalism.

Under this policy, it was advised that physicians withhold some information from the parents and child in order to achieve the best results of the child's gender acceptance. In

order to protect parents from the psychological distress of knowing the child with DSD

displayed aspects of sex and/or gender ambiguity, physicians made decisions without providing full disclosure to parents during the decision-making process.(Karkazis, 2008)

It's hard to say the extent to which some physicians omitted information and others

provided it under Money's paradigm. However, it is safe to say that the ideology of medical paternalism was inherent in the ideology of Money's approach.

Shifting ideals from medical paternalism toward evidence-based medicine and patient-centered care, and philosophical arguments about a patient's right to autonomy have caused a transition to the practice of shared decision-making. "Shared decision-making is seen as a mechanism to decrease the informational and power asymmetry between doctors and patients by increasing patients' information, sense of autonomy and/or control over treatment decisions that affect their well-being."(Eddy, 1990; E. J. Emanuel & Emanuel, 1992; Ryan, 1992 in Charles, Gafni, & Whelan, 1997) Healthcare's shift toward patient-centered care, via shared decision-making and the practice of evidence-based medicine, reflect a growing trend of patient empowerment and greater patient choice.(Department of Health, 2000)

The ideological differences between the Paternalistic Model and Shared Decision-Making Model of medical decision-making are outlined by Joosten et.al. in the following table:

<b>Table 1.</b> Models of shared decision-making about treatment [2, 42]		
	Paternalistic model (‘traditional medical model’)	Shared decision-making
Role of the clinician	<i>Active:</i> Reports only selected information to the patient, chooses the therapy he considers best for the patient.	<i>Active:</i> Reports all information and treatment possibilities to the patient. Can recommend an option. Decides on the therapy together with the patient.
Role of the patient	<i>Passive:</i> Accepts the proposal of the clinician. Is obliged to cooperate in his recovery.	<i>Active:</i> Receives all information. Forms his own judgement on harms and benefits of treatment options. Discusses his preferences with the clinician. Decides on the therapy together with the clinician.
Information	<i>One way (largely):</i> Clinician → patient	<i>Two way:</i> Patient ↔ clinician
Deliberation	Clinician alone or with other clinicians	Clinician and patient (plus potential others)
Who decides?	Clinician	Clinician and patient

(Joosten et al., 2008)

### SHARED DECISION-MAKING

Shared decision-making is a collaborative process that allows patients and their providers to make healthcare decisions together, taking into account the best scientific evidence available, as well as the patient’s values and preferences.(Wexler, 2012)

According to Charles et al. definition, Shared Decision Making includes these essential elements: (1.) there is a two way exchange of information between patient and physician including medical and personal information throughout all stages of decision-making – i.e. information exchange and analysis, values clarification, decision deliberation and consensus, implementation, and decision review(Wexler, 2012) (and in instances when multiple physicians and/or multiple members of the patient’s family or friends are also involved in decision-making, they too play a role), (2.) the possible interventions and outcomes are discussed and deliberated, and (3.) together the patient and doctor arrive at a consensus about what to do.(Charles, Gafni, & Whelan, 1999)

Organizations such as the U.S. Department of Health and Human Services, the Institute of Medicine (IOM), the Agency for Healthcare Research and Quality (AHRQ), the American College of Physicians (ACP), the American College of Surgeons (ACS), the American Academy of Family Physicians (AAFP), the Society of Critical Care Medicine (SCCM), and the American Academy of Pediatrics (AAP) have endorsed a shared decision-making approach to medical decision-making and encouraged doctors to adopt its practices. If we return our thoughts to the Hippocratic Oath, we see another way in which medicine has dedicated itself to shared decision-making. The modern version of the Hippocratic Oath used in many medical schools today was written by Louis Lasagna, the Academic Dean of the School of Medicine at Tufts University, and demonstrates a change in philosophy: Physicians pledge:

*I will apply, for the benefit of the sick, all measures which are required, avoiding those twin traps of overtreatment and therapeutic nihilism.*

*I will remember that there is art to medicine as well as science, and that warmth, sympathy, and understanding may outweigh the surgeon's knife or the chemist's drug.*

In doing so, they make a dedication to take the patient's perspective into account. No longer is the physician's word the only and final word. Inherent in the acts of warmth, sympathy and understanding is a commitment to honor the patients' perspective, including their preferences and desires. Shared decision-making is now considered the standard of care for most every medical decision<sup>17</sup> and is considered especially valuable in the face of complex or difficult medical decisions that are not consensus-based, and thus require

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<sup>17</sup> Exceptions to this process include instances when the patient does not have the capacity to participate in decision-making (as discussed in Chapter 2) or in cases of medical emergency. (Devettere, 2010)

patients to make decisions based upon person preferences and values.<sup>18</sup>(Barratt, 2008; Breitsameter, 2010b; Fraenkel & Peters, 2009)

Empirical evidence suggests that giving patients information and involving them in decision-making by including their personal understandings can result in beneficial psychological and physical outcomes. For example, shared decision-making can enhance patient satisfaction, adherence to treatment plans, confidence in health care recommendations, psychological adjustment to illness, and symptom resolution. (S. Ford et al., 2003; Fraenkel & Peters, 2009; Greenfield et al., 1988; Joosten et al., 2008; Kaplan et al., 1989; Loh et al., 2007; F. Légaré et al., 2010; Ward et al., 2003) Additionally, increased patient involvement shows a decrease the patient's willingness to accept and participate in risky treatment options(Fraenkel & Peters, 2009) and lowers the demand for some surgical treatments.(F. Légaré et al., 2010) Further evidence suggests that allowing patients to choose their medical treatment can enhance psychological well-being and quality of life.(Charles et al., 1997; Joosten et al., 2008)

There are few systematic studies of how early interventions and interactions between healthcare providers and the family affect the quality of life of persons with DSD or their parents.(Frader et al., 2004; Zeiler & Wickstrom, 2009) However, the positive effects that shared decision-making has had on patient coping, understanding, compliance, satisfaction, and quality of life have great implications for DSD management. Studies suggest that parents find these early decisions to be stressful and difficult to navigate.(Crissman et al., 2011; Duguid et al., 2007; Fedele et al., 2010; Jürgensen et al., 2006; Lev, 2006; Malmqvist & Zeiler, 2010; Sanders et al., 2011) In fact, one study even

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<sup>18</sup> What a perfect fit for DSD!

found that 19% and 13%, respectively, of parents of children with DSD reported clinically significant parenting stress and diminished adaptive coping capacity.(Duguid et al., 2007) Parents of children with DSD could benefit greatly from the implementation of a shared decision-making approach, and subsequently the children themselves could benefit, as parenting stress and coping patterns are predictive of children’s long-term emotional, behavioral, and social adjustment.”(Abidin, 1995; Colletti, Wolfe-Christensen, Carpentier, & et. al., 2008; Holmbeck, Johnson, Wills, & et. al., 2002)

#### DECISION-AIDS: FACILITATING SHARED DECISION-MAKING

Decision-aids are tools often used to facilitate the shared decision-making process. These tools are “mechanisms or interventions that have been developed to improve communication between health professionals and patients, and to help involve patients in making decisions regarding their health care.”(Agency for Healthcare Research and Quality, 2002) As presented by Elwyn et. al.:

Decision-aids “help people think about choices they face; they describe where and why choice exists; they provide information about options, including (where reasonable) the option of taking no action. These interventions help people to deliberate, independently or in collaboration with others, about options by considering relevant attributes; they encourage people to forecast how they might feel about short-, intermediate- and long-term outcomes which have relevant consequences, in ways which help the process of constructing preferences, beliefs and values leading to eventual decision-making appropriate to their individual situation.”(Elwyn, Frosch, Volandes, Edwards, & Montori, 2010)

Patient decision-aids do *not* advise people to choose one option over another. And they do *not* replace counseling from healthcare practitioners. By providing material that gives both practitioner and patient common ground, decision-aids make patients informed partners in

their own care and prepare people to discuss intervention options with their healthcare practitioners.(Steering Committee, 2005)

Decision-aids come in a variety of formats – such as decision boards, interactive videodiscs, interactive computer programs, audio-guided workbooks, pamphlets, or group presentations (O'Connor et al., 1999) – and have been created for a plethora of conditions and topics, including back care, breast cancer, cardiovascular disease, chronic conditions (like chronic pain, diabetes, lower back pain, knee or hip pain, and coronary heart disease), end of life, general health, geriatrics, mental health and depression, ophthalmology, orthopedic (like osteoporosis), prostate disorders, pain management, screening and testing, sleep disorders, weight loss, and women's health.(Informed Medical Decisions Foundation, 2012a)

Today, there are over 500 patient decision-aids available or being developed by many different individuals and groups around the world.(Steering Committee, 2005) Some, but not many, of these decision-aids have been made to address pediatric conditions, such as childhood immunizations(Wroe, Turner, & Owens, 2005), specific drug treatments(Alfaleh, Al Luwaimi, Alkharfi, & Al-Alaiyan, 2011), treatments for ear infections(Ottawa Hospital Research Institute, 2011), circumcisions(O'Connor et al., 1999), cochlear implants(Ottawa Hospital Research Institute, 2011), and decisions regarding interventions after a child has sustained a serious head injury(Fabbri, Servadei, Marchesini, Raggi, & Vandelli, 2011). Considering that decisions (and the ethics behind them) become incredibly more complex when they are being made by someone other than the patient themselves, that interventions for pediatrics generally follow the development of adult interventions, and that the use of decision-aids in medical decision-making is a fairly new

process, it is not surprising that there is a paucity of pediatric decision-aids. The additional stigma, lack of public focus on DSD, and general paucity of information relevant to DSD care means that the development of a DSD-specific decision-aid is a low priority in pediatric decision-aid creation.

A review of the efficacy of these patient decision-aids, performed by an international group of researchers known as the 'Cochrane Review Team of Patient Decision Aids,' has compiled decision-aids studies between the years of 2006 to 2009 and summarized their results. The updated review of 86 studies published in 2011 found that when patients use decision aids they: a.) improve their knowledge of the options; b.) have more accurate expectations of possible benefits and harms; c.) participate more in decision making; d.) experience more positive patient-practitioner communication; e.) select choices that are more consistent with their informed values (when the decision-aid includes values-clarification exercises); and f.) are less likely to choose elective surgery interventions (in cases where elective surgery is an option) after they consider other options.(D Stacey et al., 2011) Decision-aids have a variable effect on a patient's actual choices(D Stacey et al., 2011). Access to more information can surely introduce new options that patients may find preferable, but studies have shown that decision-aids rarely change patient's decision predispositions.(O'Connor et al., 1999) Patient decision-aids ultimately help patients become better informed about intervention options, determine their personal values, and become more comfortable with, and confident in, the decisions that they make. There are no apparent adverse effects on health outcomes or satisfaction due to the use of a decision-aid in medical decision-making.(D Stacey et al., 2011)



Another randomized control review of shared decision-making found positive longer-term effects on outcome measures. From this, the study concluded that shared decision-making is particularly suitable for long-term decisions, especially in the context of a chronic illness, and when the intervention contains more than one encounter.(Joosten et al., 2008) This finding is particularly applicable to DSD because these conditions do not disappear after a child receives a gender assignment or even after genital surgery(s) meant to “normalize” the appearance of the child’s genitals. DSD are not chronic illnesses,<sup>19</sup> but they are life-long conditions and their management often involves multiple physicians over the course of a person’s lifetime.

It is clear that decision-aids facilitate the shared decision-making process in positive and productive ways. If a decision-aid tailored to parents of children with DSD brought about even *one* of these effects listed above, DSD care would be greatly improved. Simply because a decision-aid produced successful in one patient population doesn’t necessarily mean the same will happen in another. We cannot assume linear causal chains. My response: You never know, until you try. The potential benefits that could come to thousands of parents and children with DSD (and ultimately their children) each year is well worth the investment necessary to produce a DSD-specific decision-aid.

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<sup>19</sup> Congenital adrenal hyperplasia (CAH) is the exception to this statement. People with CAH lack an enzyme needed by the adrenal gland to make the hormones cortisol and aldosterone. Severe cases of CAH known as salt-wasting CAH will cause adrenal crisis if not properly treated by taking a form of cortisol (dexamethasone, fludrocortisone, or hydrocortisone) every day.(A.D.A.M. Inc., 2012)

## POTENTIAL DSD-SPECIFIC BENEFITS TO USING A DECISION-AID

In addition to the positive effects of decision-aid use mentioned above, there are a few additional reasons why (1.) this patient/parent population could particularly benefit from the creation of a decision-aid and (2.) this patient/parent population is particularly well suited for the use of a decision-aid. They are as follows:

A DSD-specific decision-aid created with a careful attention to informational content and delivery can help medical practitioners meet legal and ethical standards for informed consent. As discussed in Chapter 2, these standards generally require physicians to provide all information a reasonable patient (or in the case of DSD, a *reasonable parent*) would find material in making an informed decision. Adults with DSD, advocacy groups, scholars, and some practitioners have questioned the content and quality of the information provided to parents making these early medical decisions for their child. In particular, they have questioned parents' knowledge of, and accessibility to, information regarding value-based decisions and the option of non-intervention in those situations. In the context of early cosmetic genital surgery, some groups have argued that, at present, parents are unable to give informed consent to these interventions because current information exchange precludes full disclosure of key information vital to make an informed, voluntary, and competent decision. (Lareau, 2003) In the parent interviews conducted by the University of Michigan research group, parents often commented that physicians did not provide them with enough information:

*I mean, people did give us pamphlets and they kind of explained stuff over and over but you really want comprehensive information. And it's hard.*

It's reasonable that parents want more information than just what was provided by medical professionals. All parents, including those of children with DSD, want their

children to grow up to be happy, and parents want information that speaks to this desire in relation to their child's condition. That is, parents desire information about their child's future quality of life – i.e. falling in love, dating, attraction, ability to develop intimate relationships, sexual functioning, and the opportunity to marry and raise children, etc.(Lee et al., 2006) – in addition to the medical information regarding their child's condition.

Once again, we return to discussions of standpoint epistemology. Since medical professionals give parents information that is deemed credible by the medical profession, and this data source is often restricted to publications in medical journals, then the information provided to parents is biased by this medical-professional lens.<sup>20</sup>(Breitsameter, 2010a; Hester, 2004) A decision-aid that – to use the parent's words – provides *comprehensive* information to parents of children with DSD would include a full account of all available DSD interventions (including non-intervention); it would include information about the quality of life experienced by individuals with DSD from reliable sources; it would provide quality information from a variety of sources and a variety of disciplines. A decision-aid created in this way would provide parents with all the information a reasonable parent would want and promote truly informed decision-making.

A decision-aid can also help parents discern what *specific* information or other extra-medical information is important to them as a unique *subjective* parent. As a

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<sup>20</sup> Informational biases are present in every discipline. Every area of study has a specific framework that focuses on certain kinds of data over others. This is not necessarily a bad thing, as every discipline needs to develop a common rhetoric for progress to be made in the field. However, practitioners need to be aware of these biases when presenting information to others outside their specialty. This is one more reason children with DSD should be treated by multidisciplinary teams: parents' need access to psychological support from a trained psychologist used in conversation with a surgeon's ability to perform a given surgery so as not to bias parent's toward surgical intervention by mere presentation of a single, biased perspective.

practitioner, you can't know what specific information parents want on an individual level unless you ask them. As a parent, you might not know what specific information you might desire unless you are presented with a list of things you could potentially want to know. For example, a list of questions commonly asked by parents to their child's healthcare providers presented in the decision-aid could (1.) act as a tool used by healthcare providers to discern parents' individualized questions, and (2.) present parents with a list of information that they could potentially want to know more about. A decision-aid could provide a standardized way of incorporating individualized patient-centered care to parents of children with DSD.

Physicians must be able to discern parents' informational needs and decision-making preferences. This becomes particularly difficult because informational needs and decision-making preferences are not stagnant and can change from person to person and decision to decision. Sometimes parents prefer to be very involved in the decision making process, other times they prefer to defer the decisions to healthcare providers. It is important to remember that the latter of these two decisions is still a valid decision, and just because a parent may not want to have the final word on a given decision doesn't mean that they do not want all relevant information explained to them. In other words, decisions about autonomy in the decision-making process do not necessarily correlate with the desire for information.(Beisecker & Beisecker, 1990; Butow, Maclean, Dunn, Tattersall, & Boyer, 1997; Ende, Kazis, Ash, & et. al., 1989; Feldman-Steward, Brundage, McConnell, & MacKillop, 2000; Siminoff & Step, 2005) Similarly, just because a parent chooses to rely more heavily on a physician's recommendation for gender assignment, does not necessarily mean that they do not want to be the primary decision-maker when it comes to decisions

about early cosmetic genital surgery. As a whole, practitioners tend to misjudge patients' and/or parents' desire and ability to be involved in the decision making process.(F. Légaré et al., 2010) This is particularly troublesome in the case of multi-decision interventions, like that of DSD care, where elucidation of these preferences is necessary for quality care. A decision-aid can help practitioners discern parents' decision-making preferences in each individual decision.

The ability of decision-aids to tailor themselves to individualized care is one of their most beneficial aspects. This ability becomes particularly important when parents are faced with value-based decisions – i.e. there is more than one reasonable option for interventions. Clinical use of preference-sensitive care is most suited to improve the decision-making process in these value-based decisions where (1.) the evidence for the superiority of one intervention over another is not available or lacking, (2.) the potential benefits and risks of each intervention are closely matched, variable, or unknown so that there is no clear “best choice,” (3.) interventions have major differences in terms of risks or side effects so that there is no clear “best choice,” and (4.) each choice has both advantages and disadvantages that require patients to make trade-offs between outcomes.(Elwyn et al., 2010; Karkazis et al., 2010; Siminoff & Step, 2005; Steering Committee, 2005) In these value-based decisions, the best choice involves matching personal values, situational beliefs and the features that matter most to a person with the option that has these features.(Steering Committee, 2005)

In these value-based situations, even given the same diagnosis people could reasonably decide on different treatments or interventions. An example of this specific to DSD was described in a clinical case study done by Jürgensen et. al.: A family had two

children both born with 17 $\beta$ -Hydroxysteroid-dehydrogenase-3 Deficiency. Children with 17 $\beta$ -hydroxysteroid-dehydrogenase-3 (17 $\beta$ -HSD-3) deficiency have a defect of testosterone biosynthesis with subsequent diminished virilization in XY individuals – some are raised as girls and some as boys. With the diagnostic information provided to parents, along with their own interpretation of their children’s physical appearance, the parents decided to raise their first child as a boy, and their second child as a girl.(Jürgensen et al., 2006) As this case report shows, diagnosis does not always imply a clear course of action when it comes to medical interventions. Only after parents discern their values in relation to each intervention in their specific case, can they rank options and make decisions about the best course of action. A decision-aid tailored to expose these value preferences can guide parents and practitioners through the decision-making process.

Finally, the use of a decision-aid for this patient/parent population could also help parents cope with their child’s condition. In most cases, DSD are not life threatening and interventions and decisions have the luxury of time on their side. When parents are waiting on test results that provide information relevant to the decision of gender assignment, for example, they may not describe the time it takes for those tests to come back as a “luxury.” Parents have expressed the stress and anxiety they feel surrounding the uncertainty of their child’s future.(Crissman et al., 2011; Duguid et al., 2007; Fedele et al., 2010; Jürgensen et al., 2006; Malmqvist & Zeiler, 2010) To them, these decisions can feel urgent, and sometimes they don’t conceptualize the fact that any decision exists at all:

*We wanted him normal, you know, as normal as possible, and it was never even a choice for us. We never discussed it, you know, not doing it.*

*It was never any question whether he was gonna go through the surgery or not.*

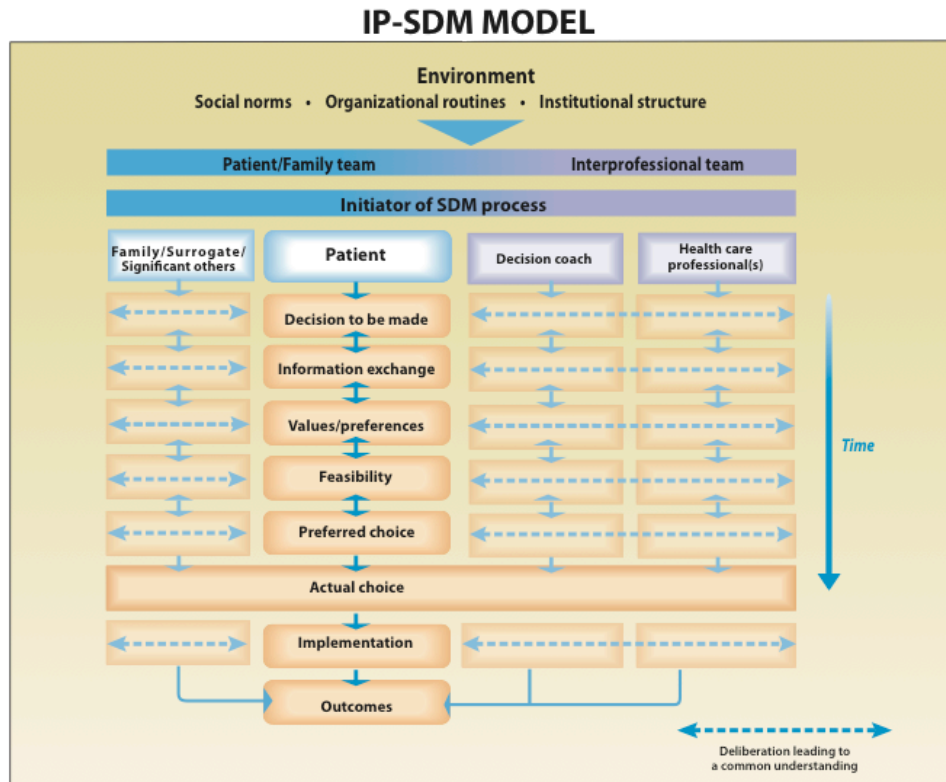
Healthcare providers must address parental stress, understanding that parents may feel as though they are in a state of emergency, even when they are not. The process of shared decision-making, including the use of a decision-aid, can help healthcare providers on the DSD team contextualize the medical facts within the parent's experience.(Monteverde, 2009) By elucidating parents' perspectives healthcare providers – including a trained psychologist – can help parents cope with the stress of the situation in ways that may not involve immediate medical intervention on the child. From a health-perspective, many of the decisions are not time-sensitive in the case of DSD interventions. The trick is helping parents come to understand this fact: There are decisions to be made, and they have time to make them.

If nothing else, a decision-aid takes time to use. In fact, current evidence suggests this is one of the reasons shared decision-making has not yet been widely adopted by health professionals. In a review study on shared decision-making the vast majority of participants (n = 2784) were physicians (89%). The three most often cited barriers to shared decision-making reported by physicians were (1.) time constraints, (2.) lack of applicability due to patient characteristics, and (3.) lack of applicability due to the clinical situation.(Gravel, Légaré, & Graham, 2006) If a decision-aid were incorporated into the standard of care for DSD patients, the time it takes for parents to go through the process of shared decision-making may help decrease parents' sense of emergency,(F Légaré et al., 2010) and increase their understanding of their child's condition and the available treatment options.(Chappuy et al., 2006) Conversely, the lack of time-sensitivity in many DSD decisions makes the use of a decision-aid more feasible for practical use.

Additionally, suggested best practice for DSD care includes treatment by a multidisciplinary team, this context may be better able to adapt to the time it takes to use decision-aids in the clinical encounter as multiple healthcare providers could potentially share this time. There is little to no data that exists to support this prediction. In fact, in 2006 a systematic review of shared decision-making in medical encounters identified 161 definitions of this process and summarized the key elements of the process into a shared decision-making model.(Makoul & Clayman, 2006) Neither the definitions nor the model included an inter-professional perspective. Inter-professionalism in healthcare – also referred to as “multidisciplinary care” – is a process by which professionals from different disciplines collaborate to provide an integrated and cohesive approach to patient care.(D’Amour, Ferrada-Videla, San Martin Rodriguez, & Beaulieu, 2005) As medical care continues to become more multidisciplinary, it would be timely for studies to test methods of enhancing patient involvement in decisions shared with multiple healthcare providers.(Marshall, Haywood, & Fitzpatrick, 2005)

In 2011, Légaré et. al. created a framework for shared decision-making in the context of a multidisciplinary team. A model of this framework shows how information and communication are exchanged between the patient (at the center of care) and the inter-professional team: (France Légaré, Stacey, Pouliot, et al., 2011)





F Légaré, Interprofessional Shared Decision-Making (IP-SDM) Model. © 2010. Available from [www.ohri.ca/decisionaid](http://www.ohri.ca/decisionaid).

As far as I am aware, this shared decision-making framework has not been tested in a specific patient population, and as the suggested DSD standard of care utilizes a multidisciplinary team approach, this DSD patient population may be the perfect group to use as a case study of this framework.

The availability of time present in many decisions regarding DSD care and the fact that suggested best practiced DSD care is done with a multidisciplinary team make DSD specifically well-suited to the use and development of a decision-aid. Given that this patient/parent population could also particularly benefit from the development of a DSD-specific decision-aid, it only seems necessary that one be developed.

### A COMMITMENT TO THE TASK

Shared decision-making can facilitate the decision-making process of parents of children with DSD in ways that are essential to making good healthcare decisions. Shared decision-making provides parents with clinical information and treatment options, as well as the opportunity to express their personal values, beliefs and goals. The use of a decision-aid in this context could facilitate the shared decision-making process in many positive and productive ways. A decision-aid carefully created to provide comprehensive, unbiased information and a framework for parents and physicians to examine all alternatives (including non-intervention) would help ensure that (1.) the best interests of the child and family are served, (2.) patient care and the doctor-patient relationship are improved, (3.) satisfaction with the decision-making process is increased for both physician and parents, (4.) decisional conflict and regret are minimized, and (5.) ethical and legal requirements for informed permission are met.(Karkazis et al., 2010) The potential benefits a decision-aid tailored to the early decisions required of parents of children with DSD are immense. Does it not seem necessary to explore if this potentiality is a reality? While the creation of a fully developed decision-aid would be an enormous task, I will take the first steps toward forming one in the next chapter.

- Chapter 4 -

**Taking the First Steps in the Development of a DSD-Specific Decision-Aid**

*Finally the doctors came in one day and explained that this test or another proved that she was a girl. They said that she had a disorder called partial androgen insensitivity syndrome. They tried to explain it to me. A lot of the information they gave us was oral. They did give us some pamphlets and they kind of explained things over and over, but then they were gone and you just have all this stuff to process by yourself. You really want comprehensive information and it's hard; the most stressful thing is just not being presented with clear-cut information. I'd never heard of this before. I had never talked to anybody that had ever experienced it and I kinda felt like... I was the only one ever having to deal with this.*

In this chapter I will present the first steps necessary for making a decision-aid for parents of children with DSD. To the best of my knowledge, this preliminary outline will be the first of its kind for this specific patient population, and is surely the first presented in the context of a larger discussion regarding the particular goals and needs of parental informed shared decision-making for children with DSD. The development of a decision-aid is a long process, but the outline presented in this chapter will be the first to move the process into the stage of actual production.

The Informed Medical Decisions Foundation, a group that works to advance shared decision-making through research, policy, clinical models and the development of a number of different patient decision-aids, published an outline of how the foundation develops patient decision-aids. The first stage of decision-aid development involves identifying the necessary content. This is done by performing a review of clinical evidence and patient-perspective literature, as well as conducting focus groups to allow patients (or parents) to express their suggestions for a decision-aid. The review of literature has been explored in previous chapters, and the latter has been started by a group of researchers at

the University of Michigan.<sup>21</sup> Parents' suggestions about a DSD-specific decision-aid have been acquired from a study conducted by this research group and will be presented in this chapter.

The second stage of decision-aid development is the actual production of the decision-aid. In this phase, a decision-aid prototype is made by creating a script and storyboard that outline the decision-aid's form and included content. Next, this outline is turned into a functioning decision-aid that is then reviewed by clinical experts and patient focus groups. The decision-aid is revised based upon the findings of this review, and finalized for clinical use.

In the final stage of a decision-aid's creation, it is put into clinical use for a given period of time and reviewed by the clinicians and patients that use the tool. Randomized control trials are conducted to determine the efficacy of the decision-aid and revisions can be made based upon the evaluation's findings.(Informed Medical Decisions Foundation, 2012b)

The development of a decision-aid is an extensive and sometimes expensive process that involves many people (patients and professionals), time, and resources. The DSD-specific decision-aid outline presented in this chapter is an essential first step in a decision-aid's creation. Since this is a preliminary outline, the information presented here is not the *exact* information (and in particular, the *exact* wording) that will be provided to the parents of children with DSD. The decision-aid used by parents will be written at a high-school reading level to accommodate parents from all educational and socioeconomic backgrounds. Ideally, this outline will be used by a team of professionals that specialize in

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<sup>21</sup> This study is one of the two that I have been using as a data source for parents' words. Some of the quotes presented throughout this paper have been from this study.

the various aspects of DSD care, advocacy, and education in collaboration with a team knowledgeable in decision-aid production to create the actual DSD-specific decision-aid.

#### FORMATTING CONSIDERATIONS FOR A DSD-SPECIFIC DECISION-AID

As discussed in the previous chapter, decision-aids can come in a variety of formats such as decision boards, interactive videodiscs, personal computer programs, web-based interactive programs, audio-guided workbooks, pamphlets, or group presentations.(O'Connor et al., 1999) An interactive computer-based decision-aid seems best suited for a DSD-specific decision-aid for a number of reasons.

First, parents have expressed that they would like a decision-aid that they could take home and use in their own time and space. This parent explained why:

*I think [a decision-aid] would need to be introduced to the family like "this is the diagnosis, these are the decisions that you're going to need to think about, and here's a guide to help you with that" – in whatever media format. Then the parents can take that home and just take some time to process. I mean I couldn't have looked at anything for a couple of days anyways. We were just sort of in shock. But then at their own leisure parents can start to kind of look at that stuff and then follow up with [healthcare providers] again after having used it and sort of processed everything.*

Another parent said:

*I think it would have been nice at that time to have had information that we could have taken with us and looked through at home when we were outside of the NICU and in a more relaxed environment.*

According to the 2000 government census, more than half of households in the United States have computers.<sup>22</sup>(Newburger, 2001) Providing parents with a tool they can use at

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<sup>22</sup> It should not be assumed that parents have a computer in their home. This is something that would need to be discussed with the family when they were introduced to the decision-aid. Appropriate accommodations should be provided to parents by the institution of care in the situation that parents do not have access to a computer.

home will ensure that parents have access to the information provided in the decision-aid whenever they want or need it.

Second, parents desire comprehensive information that addresses *all* aspects of their child's DSD. When some parents request information, they are actually expressing a desire for clear-cut, easy answers. In the world of DSD, these clear-cut easy answers are hard to come by (especially when it comes to value-based decisions) and their frequency does not increase in proportion to the amount of information provided. But parents' desire for more information cannot be dismissed. Studies have shown that parents desire more explicit information about their child's condition and the implications it will have on their child's life.(Crissman et al., 2011; Duguid et al., 2007)

*After [our child with DSD] was born we did get some information. I mean, people did give us pamphlets and they kind of explained stuff over and over but you really want comprehensive information. And it's hard.*

Parents don't just want to know the definition of DSD and their child's diagnosis. They also want to know what this DSD means for their child's life. And as this information plays a role in decision-making about the care provided to their child, their request for this information is appropriate and should be taken seriously so as to uphold the process of informed consent.

Third, it is common for people to turn to the Internet to gather their own information about their health. No matter what information the healthcare practitioners provide in the clinical setting, it is likely that parents will do additional research about their child's condition on the Internet. A decision-aid that is interfaced with the Internet could link parents to sites that contained well-rounded, accurate information presented in

appropriate ways. This connection could help deter situations like this one described by a parent of a child with OEIS<sup>23</sup>:

*[The information I received from my child's healthcare providers] was all oral. I don't remember ever getting any paper work or anything about his diagnosis. The stuff I found was all online, which was kind of bad. That needs to not be a parent's only resource because when I went online for my son's diagnosis the first site that I kept going to had autopsy pictures of these babies. That's the first thing you find when you search OEIS. It was a very horrible time in my life, very dark time. You get this diagnosis, we're told we're gonna go to [hospital 2], and so of course I go home and I try to look something up on my son's diagnosis and I'm finding autopsy pictures and recommendations to terminate the pregnancy.*

Fourth, a computer-based decision-aid can become personalized by including a section for information specific to the given parent and child. Personalization is a necessary component of DSD care because of the variability and diversity that exist in DSD cases. A paper-based decision-aid would have a much harder time making this personalized accommodation.

Finally, a study done by Protheroe et. al. found that computerized decision-aids increase patient involvement in the decision-making process, knowledge of the condition and patient's reported quality of life, while decreasing patient's decisional conflict.(Protheroe, Bower, Chew-Graham, Peters, & Fahey, 2007) For this reason, and those presented above, it seems particularly fitting that a decision-aid for parents of children with DSD be a computer-based decision-aid that can interface with the Internet. The outline presented below should be conceptualized as a series of screenshots from this computer-based decision-aid.

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<sup>23</sup> OEIS refers to the combined occurrence of Omphalocele (abdominal wall defect with gut herniation), bladder Exstrophy/cloacal Exstrophy (herniation of the bladder through the anterior abdominal wall with range in severity), Imperforate anus (the opening of the anus is missing or blocked), and Spinal anomalies.

## DECISION-AID FOR PARENTS OF CHILDREN WITH DSD: A FIRST LOOK

The shared decision-making framework for a multidisciplinary team, presented in Chapter 3, suggested that a “Decision Coach” – defined as a “health professional trained to support the patient’s involvement in healthcare decision-making but who does not make the decision for the patient” (Dawn Stacey, Légaré, Pouliot, Kryworuchko, & Dunn, 2010) – act as an interface between the multidisciplinary healthcare team and the decision-maker. (France Légaré, Stacey, Pouliot, et al., 2011) Both healthcare providers for children with DSD and the parents of these children have said that a person in this kind of role would be ideal. As this urologist said:

*I think in a perfect world, I would love to see a dedicated team that could act as liaisons between the different services.*

This parent echoed the thoughts of the physician:

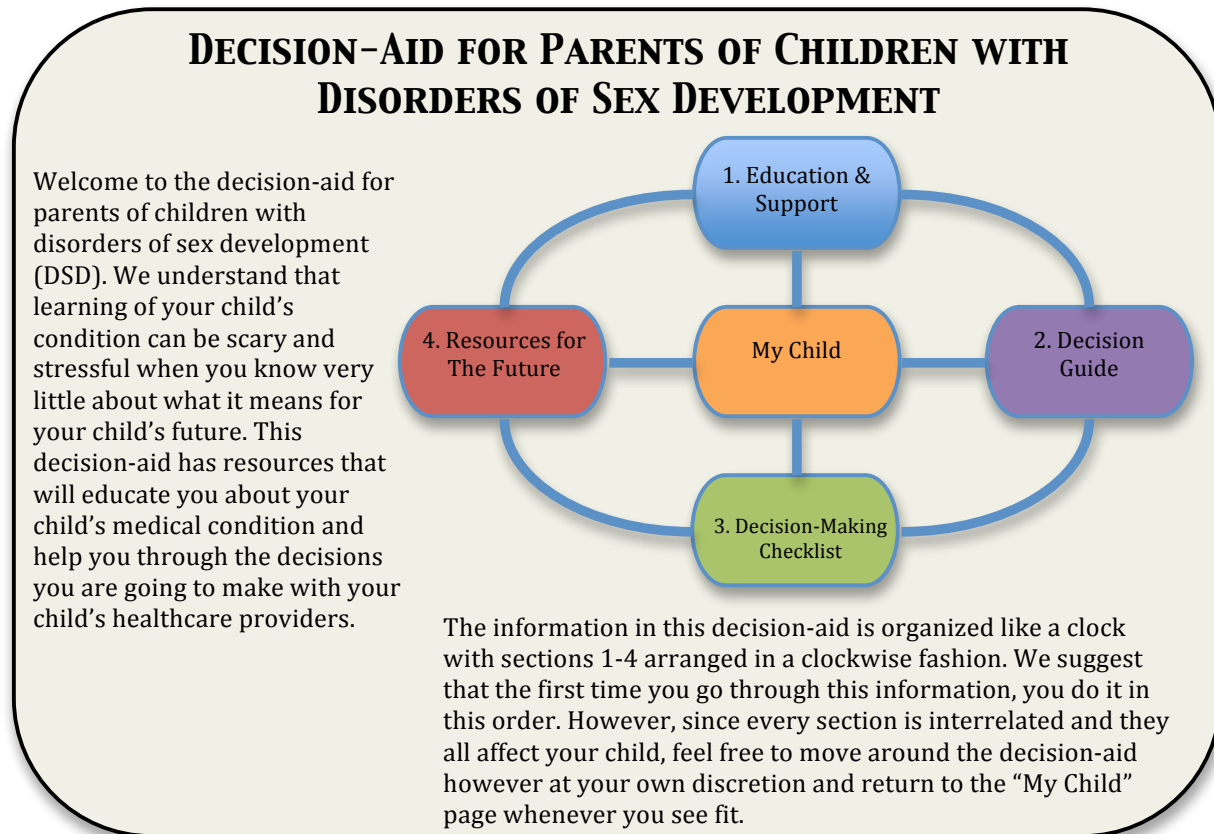
*It would have been great if there was A PERSON who was assigned to the team basically and that dealt specifically with, I mean I realize that there’s probably not enough children like ours to justify a person specifically for that but if there could be one point person who was kept in the loop with the doctors of what was going on - THAT would be perfect.*

The decision coach could act as this liaison and also be parents’ guide to using the decision-aid. Of course, parents would have access to all of the practitioners on their child’s healthcare team and have discussions with all of them throughout the shared decision-making process. However, the decision coach could be more accessible to parents and be specially trained to explain the decision-aid to them. After parents learn that their child has a DSD, the decision coach would be the one to introduce the decision-aid, teach them how to access it, and how to use it. A pamphlet that outlines this information could be provided to parents by the decision



coach so that the parents can references how to use the tool at home. An outline of this decision-aid and the reasoning behind its components are as follows.

**Home Screen: Decision-Aid for Parents of Children with DSD**



The goals of this decision-aid are to: (1.) Provide parents with facts about the medical condition, the available intervention options and their features; (2.) Help parents clarify their beliefs and values in relation to the available options; and (3.) Help parents share their values with their child's healthcare providers and others so that a course of action can be planned. To achieve these goals, the decision-aid was divided into five sections: (a.) "Education & Support" will address the informational needs of parents so that they can make informed decisions regarding their child's care. (b.) The "My Child" section is meant to foster patient-centered care by holistically grounding the decision-aid on the

specific situation of the child. (c.) The “Decision Guide” section is meant to elucidate parent’s personal preferences, values, and beliefs in relation to the decisions they must make regarding their child’s care. (d.) The “Decision-Making Checklist” is an outline of the informed shared decision making process and can be used as a check to ensure that this process has occurred. Finally, (e.) “Resources for the Future,” includes tools to address the continued care of children with DSD as they grow.

### **Section1: Education & Support**

## EDUCATION & SUPPORT

If specific information in this section seems of value to you, or if you have a question about it and would like to flag it for later discussion, you are able to “bookmark it.” The information will be transferred to your *My Child* page and you can discuss this information with your child’s healthcare providers at your next appointment.

You are not alone.  
Many parents of children with DSD have been faced with the same decisions that you will make for your child. Below there is a link to what other parents have considered and felt throughout this decision-making experience. Since every child’s situation is different, these parents cannot tell you the answers to the decisions you will make for *your* child. They can, however, provide examples of how to approach these decisions. There is also a link to what some adults with DSD have to say about their lives. Lastly, there is a link that outlines the reasons why there is controversy over some DSD interventions.

- [Parents’ Words](#)
- [People living with DSD](#)
- [DSD Controversy](#)

Education

Disorders of Sex Development

Sex Development

Sex, Gender & Sexuality

Index & Definitions

FAQ

Additional Resources

Peer Support: Words from the Wise

The informational section of this decision-aid is comprised of two modules: “education” and “support.” Together, the two modules provide the comprehensive information parents need to make informed decisions. The first module, “education,” provides parents with information about DSDs and their medical management.

The “Sex Development” link is meant to inform parents about fetal sex development and the known etiologies of DSD. This section will include graphics because parents have said that visual aids greatly increase their understanding of sex development.

*Dr. [1] was a lot better than Dr. [2] because he had a much better bedside manner of explaining things. He would draw me pictures ‘cause I’m much more visual than just trying to listen to you describe everything, so he was good.*

The “Index & Definitions” link would provide a catalogue of all the different DSDs and aspects of DSD care and interventions. For example, the definition of *CAH* and *hormone replacement therapy* would be found under this link. Parents would be able to look up the definitions that relate to their child’s specific situation and “bookmark” them for further attention in the “My Child” section.

The educational section would also include definitions of sex, gender, and sexuality and the ways that they intertwine, but don’t always normatively align. As discussed in Chapter 2, parents and practitioners have experienced discomfort discussing these topics because they are generally considered to be private. By putting this “Sex, Gender & Sexuality” link into the decision-aid, practitioners can be assured that this information is accurately presented to parents while they are in the comfort of their own homes (i.e. the private domain), thus promoting a comfortable, foundational understanding of the terms. Of course, additional conversations will occur between parents and their child’s healthcare providers regarding these topics, but introducing this information to parents in *this* way may help to decrease the discomfort experienced in these conversations.

The “Frequently Asked Questions” (FAQ) link would provide lists of questions commonly asked by parents to their child’s healthcare providers and lists of questions that parents *should* ask their child’s providers. For example, parents have requested a list of

pros and cons of doing early surgery, and also a list of questions that parents might want to ask their child's surgeon regarding early surgery. This parent said she would have liked to know more about:

*...the cost, the surgery techniques, the controversy, how to take care of your kids after surgery, [what] to do in schools – all kinds of basic stuff.*

Lists of questions can provide a guideline for healthcare providers to discern parents' individualized questions and thus promote patient-centered care, while simultaneously providing parents with an outline of important information that they may need or want to know more about. As discussed in Chapter 3, a practitioner can't know what specific information parents want on an individual level unless they ask them (i.e. the list of questions). And a parent might not know what information they might want to know more about unless they are presented with (a list of) information that they could potentially desire.

Finally, the "Additional Resources" link would provide parents with links to other quality tools or sites on the Internet – including, for example, diagnosis-specific support groups like CARES and more general informational support sites like Accord Alliance. The consensus statement encourages this collaboration between healthcare professionals and support groups (Lee et al., 2006) saying that "parents need to be informed about sexual development" and DSDs, and "Web-based information may be helpful, provided the content and focus of the information is balanced and sound." (Lee et al., 2006) The "Additional Resources" link would provide parents with links to quality information from a variety of sources. This parent said that having information consolidated in this way would be beneficial:

*I think the one thing I've really struggled with – as far as the information that is out there – is that I have to pick and choose from different resources to get it. It's like there's so much information out there and it needs to be all put in one place...*

The second module in this informational section is “Support.” This module provides parents with first-person vignettes of other parents of children with DSD and also adults living with a DSD. As displayed in this parent’s words, some parents feel very isolated when they first learn of their child’s condition:

*In the beginning I didn't know anything about [his DSD]. I had never talked to anybody that had ever experienced it, and I kinda felt like I was the only one ever having to deal with this and that – that you know [my child] was different.*

The purpose of this module is to break down that feeling of isolation by showing parents that others have been in their position (and their child’s situation) before. The “Parents’ Words” link will provide first-person accounts of parents’ early experiences and decision-making *approaches*. Parental vignettes will not include the actual decisions that they made but rather showcase a variety of decision-making styles. Parents could use these styles to help them identify what approach they most emulate, or would like to emulate in the decision-making process. The “People living with DSD” link will include short first-person accounts of what life with a DSD is like. Their words will emphasize the fact that individuals with DSD have the potential to become happy, well-adjusted, functional, members of society (Lee et al., 2006) – which is an important piece of information when it comes to comprehensive care. Again, these first-person accounts will not be presented as outcome case studies for different intervention decisions. Rather, they will be used to emphasize person-first philosophy: People with DSD are not defined by their condition; their condition is just one of many things that make them who they are.

The final link in this module, “DSD Controversy,” will explain the difference between the consensus-based and value-based decisions parents will be making for their child. An outline of the variability in intervention decisions will be presented as well as an outline of the particular trade-offs that are inherent in choosing one decision over another. Parents will be reminded that physicians will not always be able to tell them which decision is in their child’s best interest, and that in these situations the decision is dependent on what parents believe is right for their child.

The comprehensive information provided in this section is meant to educate parents about the general aspects of DSD so that they can come into conversations with their child’s healthcare providers feeling prepared. The fact that the information comes from a variety of sources and perspectives will decrease standpoint biases from any one position. This section is meant to provide parents with the information breadth, not depth. Since every child’s condition is different and every family is different, this educational section cannot be used to replace detailed, personalized, consultation with healthcare providers necessary for informed decision-making.

**The Center of Care: My Child**


## MY CHILD

We all have visions of what our child's future will hold. This section is meant to help you think about what you want for your child's future and how your child's DSD will actually impact their ability to achieve these goals.

Goals of Life Exercise

The "My Notes" section is a place where you can write down any information for yourself.

My Notes



The "Important Information" section is a list of all the information you selected in the "Information & Support" section of this decision-aid.

My Important Information

Part of providing patient-centered care is providing individualized "whole person" care.(Bechtel & Ness, 2010) This section of the decision-aid is meant to highlight this focus on the child. The section allows parents to review the information they "bookmarked" in the *Education & Support* section because of its importance to their child's situation. This section also provides a space for parents to write in their own notes. e.g. "Remember to ask Dr. X about Y," or "Z is a good resource," thus promoting individualized attention to *this* child's situation. To support "whole person" care, physical pathology needs to be addressed in the larger context of the person's life. The Consensus Statement suggests that the psychosocial management of children with DSD should include discussions with parents regarding the child's future quality of life. The "Goals of Life Exercise" would be a tool used to promote the discussion of this individualized, whole person approach to care.

When asked what parents want for their child, they say things like, “I just want my child to be happy.” The “Goals of Life Exercise” would ask parents what this entailed for their child’s future. Does it mean that their child does well in school? That they play sports? Does it mean that they have friends and, in the future, a loving partner, or perhaps children some day? These “life goals” as we call them are ideals that most parents wish for their children. In this exercise, parents would be able to express the life goals and future ambitions they have for their new baby.

Parents would then be reminded that “falling in love, dating, attraction, ability to develop intimate relationships, sexual functioning, and the opportunity to marry and raise children,” are all achievable goals for their child’s future, regardless of their child’s biological indicators of sex.(Lee et al., 2006) Achieving these life goals is easier for some people with DSD than others, and support should be given to those who need it. Sexual aversion and lack of sexual arousal, (which are often misinterpreted as low libido)(Basson, Leiblum, Brotto, & et. al., 2003), or avoidance of intimate relationships in fear of rejection, are three of the most common hardships faced by people with DSD.(Lee et al., 2006) Parents need to be aware of these issues, but reminded that all of these hardships are faced by countless other people in the world that don’t have DSDs.

As humans, we have a tendency to focus on what’s different, not what’s the same. This bias is known as the “focusing illusion” and it occurs when people pay too much attention to certain details while largely ignoring other (equally important) factors.(Wasko & Pury, 2009) Since the things we tend to focus on are the factors that are more alarming or have more worrisome effects, and these factors are often judged more likely to occur, this can cause serious biases in the decision-making process.(Rector, 2008) Parents have



expressed a great deal of stress about their child's future after learning of their child's DSD, and often exaggerate the affect of the condition on their child's future quality of life. For those children with atypical somatic sex development, it's easy for parents to start focusing on how their child's genitals make them different from other children, and forget the bigger picture of how they are the same. Since we tend to give more weight to the things that we focus on, this attention to the genitals can distort parent's decision-making process by exaggerating the effects atypical genitalia will have on the child's quality of life.(Kahneman & Thaler, 2006)

The focusing illusion is one of the biases that influence *affective forecasting*. Affective forecasting is a process in which someone thinks of a given situation and makes a prediction about how one will feel about the situation in the future. As it turns out, people are really bad at predicting how they will feel in the future given a certain situation.(Halpern & Arnold, 2008; Tversky & Kahneman, 1974) This has serious implications for medical decision-making as many health decisions depend on the ability of the patient (or parent) to think about the ways specific medical interventions will affect their future and how they will feel about that. When people exaggerate the duration and intensity of a negative factor (such as atypical genitalia) on their future they could potentially make decisions that are not in accordance with the reality of that future. (Gilbert, Pinel, Wilson, Blumberg, & Wheatley, 1998; Kahneman & Thaler, 2006)

Particularly relevant to parents of children with DSD, is the finding that non-patients (e.g. parents) consistently underestimate the quality of life associated with chronic health conditions (like DSD) of others.(Halpern & Arnold, 2008; Ubel, Loewenstein, Schwarz, & Smith, 2005) Reinforced by the "Peer Support" module in the previous section, this section

presents parents with well-rounded information – including the ways in which a child with DSD has a bright future just like every other child – and can help parents resist focusing on their child’s differences while making proxy-decisions.

## **Section 2: Decision Guide**

### **DECISION GUIDE**

After you have learned what you need to know about DSD and clinical management strategies, you may have to make decisions regarding your child’s care. The decision guides (at right) are designed to assist you and your child’s team of healthcare providers in the decision-making process. The guides will help you determine which factors matter most to you in a specific decision. Reviewing these with your child’s healthcare providers can help you determine what decision best fulfills these values.

Know that the decisions you face are highly dependent on your child’s specific situation. For this reason, you may use *all* of these decision guides, you may use *some* of them, you may even use the “Make Your Own” outline as you explore your concerns and decisions.

Gender Assignment

Early Surgery

Sharing Information

Make Your Own

A main feature of the shared decision-making process is the elucidation of parent’s personal beliefs and values regarding the medical management of their child’s DSD. Intervention and non-intervention in DSD care imply a series of trade-offs where valuing one aspect could mean having to give up others. This section of the decision-aid is meant to assist parents in this process of deciding what they care about most, and subsequently foster conversations between healthcare providers and parents about what these values would imply for the child’s medical interventions. As we will see, the tools used to evaluate parent’s values serve multiple purposes. Not only do they help parents identify and assess

their own values, but these tools also work as a form of checks and balances between parents and healthcare providers.

Identifying the most common decisions parents must make that require them to assess their values is a first step in the development of these tools. The next step involves outlining all the factors that influence these decisions. Some qualitative studies on the early parental experience of making decisions for their child with DSD have suggested that the most common value-based decisions parents face are gender assignment, early cosmetic or elective surgical interventions, and whether or not to tell others about their child's condition.(Crissman et al., 2011; Jürgensen et al., 2006; Sanders et al., 2011) When the University of Michigan research group spoke with twenty parents of children with DSD about their experiences of making early decisions, parents mentioned a number of factors that they took into account for a number of considerations. The two decisions that parents most often referenced were gender assignment and early surgery. The lists of the factors that parents took into consideration while making decisions in these two domains are as follows:

<b>Factors Parents Referenced when making Decisions about...</b>	
<b>Gender Assignment</b>	<b>Surgery</b>
Genetic Testing/ Karyotype	Timing of Surgery (Pros and Cons)
Hormone Levels	Effects on potty training
Physical Appearance	Child's memory of the operation(s)
Religious Views – i.e. “We didn’t want to play God.”	Child's memory of the pain of the operation(s)
Parental Intuition/ Gut Feeling	Physical Appearance
Interpretation of Child's Actions – i.e. Gender-normative play	Preferences about child's autonomy in decision-making

Urgency and importance of any other conditions	Function – i.e. Ability to ejaculate or repair of cavity confluence
Lab test results	Form: “Normal Appearance” – i.e. the ability to pee standing up
Gonadal material and/or Development	Cost of surgery and Available surgical techniques
Comfort with Doctor and/or Institution of Care	
Fertility/ Potential Fertility	
Decision-Making Preference – i.e. “How involved do I want to be in the decision-making process?”	

No parent referenced all of these factors when discussing why and how they made the decisions that they made. Some parents would focus on a specific set of factors and other parents would focus on another set. On the one hand, this selectivity could be a sign that parents were lacking information. For if they didn't know that a given factor could even play a role in their decision, then how could they have referenced it when speaking about their decision-making process? On the other (and more likely) hand, the selectivity that parents expressed when discussing the relevant factors to a given decision could be a reflection of their value preferences. When it comes to multi-factorial decisions, parents are encouraged to use their personal beliefs and values to rank the importance of factors against each other. The factors of a decision that are ranked most important are then matched with the decision that best fulfills those factors.

*Values clarification exercises* are tools that help parents determine their preferences for a given decision. These exercises help parents identify the dimensions of the decision (i.e. all the factors that could play a role in decision-making) and also help them assess, explore, and determine what their personal values are in relation to each factor. Common values clarification exercises include ranking options or putting options of different

“weights” on a balance beam or a scale and exploring what these weights would imply about the best course of action. A prototype values clarification exercise, which can be characterized as a series of balance beams or scales, was created for parents of children with DSD making decisions about gender assignment. This model was created by taking the factors that parents mentioned above and inserting them into a model values clarification exercise created for women making decisions about the use of a breast cancer drug (Tamoxifen). (O’Connor, 1999)

<b>Values Clarification Exercise for Parents of Children with DSD Making Decisions about Gender Assignment</b>										
<b>Below is a list of factors other parents of children with DSD have considered when making a decision about their child’s gender assignment. Please indicate how important these factors are for you by circling the number from 1 (not at all important) to 10 (extremely important) in making your decision about your child’s gender assignment with your child’s healthcare providers.</b>										
Some people, after having received all the information they need about gender assignment and possible intervention options, prefer to leave decisions about gender assignment up to the doctor. Others prefer to participate in these decisions in a variety of ways. Which statement best describes what you believed is the ideal way for the decision to be made?										
_____	I prefer to leave all decisions regarding my child’s gender assignment to the <b>doctor</b> .									
_____	I prefer that the <b>doctor</b> make the final decision about gender assignment, but seriously consider my opinion.									
_____	I prefer that the <b>doctor</b> and I share responsibility for deciding which treatment would be best for my child.									
_____	I prefer to make the final decision about my child’s gender assignment after seriously considering the doctor’s opinion.									
_____	I prefer to make the final decision about my child’s gender assignment. <sup>24</sup>									
How important is the congruence between your child’s genital appearance and their gender assignment?										
1	2	3	4	5	6	7	8	9	10	
Not at all									Extremely	

<sup>24</sup> This first question referring to a parent’s decision-making preference was modeled off of a questionnaire made to determine the “Information needs and decisional preferences of women with breast cancer.” (Degner, Kristjanson, & Bowman, 1997)

Important	Important
How important is the congruence between your child's karyotype and their gender assignment?	
1 Not at all Important	2    3    4    5    6    7    8    9    10 Extremely Important
If applicable, how important are the results of any other lab tests to your child's gender assignment?	
1 Not at all Important	2    3    4    5    6    7    8    9    10 Extremely Important
How important are the effects of pre-natal hormone levels on your child's brain to your decision about your child's gender assignment?	
1 Not at all Important	2    3    4    5    6    7    8    9    10 Extremely Important
How important is the presence of gonadal material (i.e. male-typical testicular tissue or female-typical ovarian tissue) to your decision about your child's gender assignment?	
1 Not at all Important	2    3    4    5    6    7    8    9    10 Extremely Important
How important is your child's fertility or potential for fertility in your decisions about your child's gender assignment?	
1 Not at all Important	2    3    4    5    6    7    8    9    10 Extremely Important
How important is it to have a gender assignment as soon as possible?	
1 Not at all Important	2    3    4    5    6    7    8    9    10 Extremely Important
How strong of a "gut feeling" or intuition about which gender would best fit your child do you have?	
1 Not strong At all	2    3    4    5    6    7    8    9    10 Extremely Strong
If you do have a strong "gut feeling," what is it? _____	
Sometimes children with DSD have other health conditions that must be addressed first. Are there any other decisions that are more important right now, and should be addressed first?	

No _____	Yes _____
How prepared do you feel to make a choice about your child's gender assignment?	
1 Not Prepared At all	2      3      4      5      6      7      8      9      10 Extremely Prepared
How comfortable do you feel with the healthcare providers that are working with you to make decisions about your child's gender assignment?	
1 Not at all Comfortable	2      3      4      5      6      7      8      9      10 Extremely Comfortable
How comfortable do you feel with the healthcare institution in which you are making decisions about your child's gender assignment?	
1 Not at all Comfortable	2      3      4      5      6      7      8      9      10 Extremely Comfortable
Are there any other factors that play a role in decisions about your child's gender assignment? If "yes," what are they and how important are they to your decision?	
* _____	
1 Not at all Important	2      3      4      5      6      7      8      9      10 Extremely Important
* _____	
1 Not at all Important	2      3      4      5      6      7      8      9      10 Extremely Important
* _____	
1 Not at all Important	2      3      4      5      6      7      8      9      10 Extremely Important

Parental use of values clarification exercises created in this way will promote parents' critical evaluation of values relating to all the factors in a given decision – including their desired involvement in decision-making. As discussed in Chapter 2, evidence suggests that when physicians infer patient preferences for information and for participation in treatment decision-making, they are often wrong (C. Charles, Gafni, &

Whelan, 1997; Ryan, 1992; Strull, Lo, & Charles, 1984; Waitzkin, 1991) And as discussed in Chapter 3, parents' decisions about autonomy in the decision-making process do not necessarily correlate with their desire for information.(Beisecker & Beisecker, 1990; Butow et al., 1997; Ende et al., 1989; Feldman-Steward et al., 2000; Siminoff & Step, 2005) Because informational needs and decision-making preferences are not stagnant and can change from person to person and decision to decision, this first question should be included in each decision parents make. The variability in parents' decision-making preferences are showcased below:

*I think in the beginning I wish they would have given me a larger role in the decision-making process. I think after we left the hospital, I kind of told everyone what my role was going to be. Whereas, when I was in the hospital I think I felt very overwhelmed and to some degree wanted everyone to tell me what to do. Because I was just completely...I felt completely in over my head.*

*I guess knowing from my own personal experiences, it is important to be involved in your own healthcare – that you are a vital part of your own health. Doctors are there with their education, knowledge, and expertise but you can't just hand your life over to them. So I was prepared to be a part of making the decisions.*

*I would probably have to say smaller because that was a lot on the plate they handed to me.*

Values clarification exercises may also serve as a system of checks and balances on the information exchange and understanding between parents and providers: On one side, parents check to make sure they have been provided with all the relevant information to a given decision, and on the other, providers can check to make sure parents are understanding the information they are provided. For example, (using the values clarification exercise presented above) if a parent read the question, "How important are the effects of pre-natal hormone levels on your child's brain to your decision about your



child's gender assignment?", and the parent had never heard of pre-natal hormones before, then they would know that they need to ask their child's healthcare provider about this factor. By including all the relevant factors, the tool would give parents an indication that they are missing some information if they come across a question of which they had no previous knowledge.

Conversely, the values clarification exercise can help practitioners elucidate parents' understanding of the information they are provided. Using the prototype example presented above, we know that all of these factors play a role in some way, shape, or form in decisions regarding gender assignment. It is completely acceptable for parents to have preferences in as to which factors are most important to them. But since all of the factors work in conversation with each other, it is not acceptable for one factor to over-ride the decision-making process at the expense of all others.

A qualitative study of parents' early decision-making experiences found that parents tend to have simplistic associations between certain factors and the role they played in decisions about their child's gender assignment.(Crissman et al., 2011) For example, when thinking about gender assignment this parent said:

*I think it was...XX is female and XY is male...and his test came back XY showing that he was a male and even though the testicles were not descended, the test would actually let them know if they were actually testes or ovaries. So, I guess it was just a chromosome test that everything was hinging on.*

If a parent were to fill out the exercise with the weight of chromosome test results at a "10" and all other factors received a "1" – as the parent above could have done – then healthcare providers would know that additional discussions are needed to determine why parents indicated these values and to help parents understand gender assignment as a

more nuanced interplay of multiple factors. Used in this way, values clarification exercises can help practitioners and parents recognize gaps, inconsistencies, and exaggerations in parents' reasoning before they make potentially irreversible decisions for their child.

Values clarification exercises do not give parents answers to the decisions they must make. They simply help them assess their values regarding a specific decision. Once these values have been examined, parents and practitioners can determine a course of action. Ultimately, these exercises should be used as a tool to facilitate the shared decision-making process for both parents and healthcare providers by fostering meaningful conversations between the two parties. These exercises could be created for the plethora of decisions parents of children with DSD may face, including surgeries (such as early cosmetic surgery to “normalize” external genital appearance or the removal of gonadal material) and whether or not to share information about the child's DSD with others. All of these decisions have multiple factors that are intertwined with each other via a series of trade-offs and require value assessments before a decision can be made.

**Section 3: Decision-Making Checklist****DECISION-MAKING CHECKLIST**

- I understand that my child has been diagnosed with a DSD
- I understand that DSD are not life-threatening conditions (except in some cases of CAH)
- I understand that children with DSD can grow up to live happy and fulfilling lives with close friends, family, and romantic partners and the ability to do this is not dependent on the appearance of their genitals
- I have spoken with my child's healthcare providers about what is known about the physical aspects of my child's DSD, e.g. karyotype, hormone levels, potential for fertility, etc.
- I recognize that there are decisions to be made regarding the clinical care of my child's DSD – sometimes including gender assignment, early surgery, or disclosure – and that there is inherent uncertainty in the outcome of these decisions.
- I have used the decision-guides to help me organize my values about specific aspects of my child's DSD and I have spoken with my child's healthcare providers about what course of action has the features that best match my values
- I have asked all the questions that I feel I need to ask in order to make an informed decision that is in alignment with my values
- I feel prepared to make a decision

Informed Decision Document

Informed shared decision-making is a multi-stage *process*. In this process, practitioners provide the necessary clinical information parents should know and assist them in understanding that information. Parents must then evaluate the available treatment options by integrating their own values with the known facts. The two parties must discuss and deliberate about these options, and finally come to an informed shared decision about intervention or non-intervention. From a moral perspective, an informed decision will have occurred when this process has occurred, even if the informed decision document has not been signed.(Devettere, 2010) The shared decision-making checklist essentially outlines the *process* of informed shared decision-making. By walking a parent through this process, we can help ensure that parents are making informed shared decisions regarding their child's care.

Attached to this module would be a copy of an informed decision document.<sup>25</sup> The legal signing of the document is an acknowledgment that the process of informed decision has occurred.(Devettere, 2010) Currently, parents do not sign a DSD-specific informed decision document.<sup>26</sup> The informed decision-making process for parents of children with DSD is wrought with hardships. Why not acknowledge these difficulties in the document by including clauses that specifically address their implications? The informed decision documents used in clinical practice today only recognize the decision to *do* a specific intervention. These consent forms are not used to acknowledge the decision *not* to intervene. In our society it is important to have a “paper trail” for medical decision-making – especially in the face of treatment controversy and medical-legal liability. Additionally, the right of the child’s future adult-self to obtain information about the treatment(s) he or she received during childhood can only be upheld through seamless documentation of that process.(Wiesemann, Ude-Koeller, Sinnecker, & Thyen, 2010) The decision not to intervene needs to be recognized and documented as a valid decision in the same way an intervention is recognized and documented. As Devettere puts it, “Choosing to do nothing in a situation where we could do something is just as much of a choice as the choice to do

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<sup>25</sup> I use the phrase, “Informed Decision Document” instead of “Informed Consent Document” to stay consistent with the terminology I designated in Chapter 2. Use of the terminology “informed choice/informed decisions” indorses a commitment to disclose *all* treatment alternatives rather than the *one* alternative that the physician deems best, as implied by “informed consent.” (Charles et al., 1997) As far as I’m aware, these documents are still referred to as “informed consent documents” in the legal and medical setting.

<sup>26</sup> In his “General Discussion of Legal Issues Affecting Sexual Assignment of Intersex Infants Born with Ambiguous Genitalia,” Glenn Burton created a DSD-specific informed consent document specific to early cosmetic genital surgery. His document includes an outline of information a parent needs to know in order to give informed consent. However, Burton’s form does not include a space to consent to non-intervention – that is, the decision not to do surgery. Since the decision not to intervene is a decision in itself, his should be revised to include this option.

something. We are responsible both for what we freely choose to do and for what we freely choose not to do but could do.”(Devettere, 2010) As long as parents have gone through the informed shared decision-making process outlined in the checklist above, both intervention and non-intervention are informed decisions that should be recognized in the signing of a DSD-specific document.

Healthcare providers would be responsible for making sure this informed shared decision-making process has been achieved before decisions are implemented. Physicians also need to know parents have the capacity to make a given decision at a given time, and discussing the “Decision-Making Checklist” would facilitate this assessment. In addition to conversations to confirm understanding, before parents sign the informed decision document, physicians could ask them to complete a series of questionnaires to evaluate if parents are ready to make the decision at hand. The following three questionnaires could be administered to the parents to assess their readiness for making these important decisions: (1.) PRIME-MD (Mood & Anxiety Modules) Questionnaire(Spitzer et al., 1994) – this questionnaire identifies the level of anxiety and stress in parents; (2.) Decisional Conflict Questionnaire(A.M. O’Connor, 1995) – this questionnaire’s aim is to elicit three components of decisional conflict: (a.) health-care consumers’ uncertainty in making a health-related decision, (b.) the factors contributing to the uncertainty, and (c.) health-care consumers’ perceived effective decision-making. In preliminary studies, the scale was able to differentiate between people with a clear intention for a certain decision vs. those who chose to delay their decision-making; and (3.) Realistic Expectations Questionnaire (O’Connor, 1995) – this questionnaire assesses if patients understand the possible benefits and limitations of a given intervention. A series of these questionnaires would need to be

created for the plethora of decisions parents face. An example of the kind of material that could be included on a DSD-specific realistic expectations questionnaire for early genital surgery could be, “Do you understand that even after your child’s surgery is concluded, your child still has a disorder of sex development?” or “Do you understand that your child may require additional surgeries in the future to achieve the intended outcome?” In combination with the Decision-Making Checklist, these questionnaires could help ensure that parents are ready to make informed shared decisions for their child.

#### **Section 4: Resources for the Future**

### **RESOURCES FOR THE FUTURE**

As your child grows you will begin to educate them about their DSD in age-specific ways. Attached are resources that other parents of children with DSD have found helpful that you can use to help you with your child when the time comes.



Educating your Child  
About their DSD

School

Suggestions from Others

Additional Resources

This final section of the decision-aid provides parents with resources to address the future care of their child in relation to their DSD. As the child grows, parents need to inform their child about their DSD in age-specific ways. (J. Money, 1994) The Consensus Statement says that this process of educating the child about their condition – i.e. the process of

disclosure – concerning facts about karyotype, gonadal status, and prospects for future fertility is a collaborative, ongoing action that requires a flexible individual-based approach and should be planned with parents from the time of the child’s diagnosis.(Lee et al., 2006)

The “Educating Your Child about their DSD” link would provide parents with information appropriate to specific age-groups. It would also provide parents with a list of common signs children show when they are ready to receive more information. This information would be compiled with the help of psychologists that specialize in pediatric DSD cases, and the practical knowledge of parents. There are some books that other parents have found helpful to teach their child about various aspects of their DSD. This kind of tool would be found under “Additional Resources.”

Parents have also requested information about how to help their child cope with having a DSD (should that kind of intervention become necessary). Parents often worry about what would happen if other children at school found about their child’s DSD:

*I guess it would be school. You know in school when a child would see him and know he doesn’t have a penis and tell somebody else. Then that person tells another person then other people look at him differently. That’s what’s concerning to me.*

The “School” and “Suggestions from Others” links would include information from psychologists and other parents about how to help children address these issues. This information has not yet been gathered, but future stages of this decision-aid’s development could incorporate qualitative focus groups with parents and psychologists to develop prototype material.

## BEST PRACTICES, BENCHMARKS &amp; A MORAL REMINDER

This preliminary outline is a necessary first step in a decision-aid's creation specific to parents making decisions for their children with DSD. As discussed at the beginning of this chapter, this preliminary decision-aid has a long road of development before it is ready for clinical use. For argument's sake (and for the sake of parents of children with DSD), let's say that the outline presented here ultimately leads to the creation of an actual decision-aid. Implementing new protocol in medical practice is notoriously difficult; Physicians find it hard to incorporate new evidence-based findings into clinical practice.(Godin, Bélanger-Gravel, Eccles, & Grimshaw, 2008; J. M. Grimshaw, Eccles, Walker, & Thomas, 2002) and institutional barriers frequently resist changes that require a restructuring of physical space or monetary allocation.(Dreger & Sandberg, 2010) The use of a DSD-specific decision-aid would require both physicians and their institutions to make changes to the way they provide care for this population. For this decision-aid is not a brief handout physicians would distribute to a parent, but rather an interactive framework for ongoing discussions and care. And should a decision coach be utilized to act as liaison between parents and the multidisciplinary team, the hospital would have to compensate the coach within the treatment paradigm. The implementation of a DSD-specific decision-aid into clinical practice could be faced with many challenges.

For implementation to occur in a standardized, practice-wide way, a few things would need to happen. First, the decision-aid would need to be tested in randomized control trials. If the studies found that those parents who used the decision-aid made better decisions (i.e. have improved knowledge of the options, have more accurate expectations of possible benefits and harms, participate more in decision making, experience more positive



patient-practitioner communication, and select choices that are more consistent with their values) as compared to controls, adherence to evidence-based medicine would imply that the decision-aid should be used in clinical practice. Next, the use of the decision-aid would need to be incorporated into best practice standards. These standards reflect clinical management strategies or interventions that consistently show results superior to those achieved with other means, and can be a benchmark for quality care.(Bogan & English, 1994) Those institutions that meet the standards of best practice are considered to provide optimum care. Independent groups, like Accord Alliance, that make recommendations to parents of children with DSD about where they can find the best care for their child, would be able to rank healthcare teams based upon their use (or lack there) of the decision-aid (among other criteria). These standards and benchmarking of quality care would push institutions and providers to incorporate the decision-aid into their healthcare protocol for children with DSD.

Recalling the Hippocratic Oath: The moral principle of beneficence mandates that physicians “first do no harm.” A DSD Decision-aid would facilitate the physicians’ ability to maximize health benefits and minimize health risks to their patients, while providing detailed information in adherence to mandates for informed decision-making and encouraging parents to share the decision-making process which is considered standard best practice in medical decision-making.

- Conclusion -

**A Call to Action: Part II**

*Knowing is not enough, we must apply.  
Willing is not enough, we must do. – Goethe*

During times of stress and anxiety parents must make early decisions for the medical management of their child's DSD. Parents want to make the best decisions that they can for their child, but their words sometimes indicate that they are not prepared to do so. From what parents have directly said, we know that they desire for more, quality, information about their child's condition and their child's prospective future. From what parents have indirectly said in discussions about their early decision-making experience, we know that parents sometimes lack understanding of diagnostic tools and clinical findings, have unrealistic expectations for interventions, and don't even acknowledge that there are decisions to be made regarding their child's care. Particular attention to information exchange via informed decision-making, and collaborative preference-sensitive care via shared decision-making are pivotal to helping parents make the best decisions for their child.

Clinical use of preference-sensitive care is most suited to improve the decision-making process in decisions where (1.) the evidence for the superiority of one intervention over another is not available or lacking, (2.) the potential benefits and risks of each intervention are closely matched, variable, or unknown so that there is no clear "best choice," (3.) interventions have major differences in terms of risks or side effects so that there is no clear "best choice," and (4.) each choice has both advantages and disadvantages that require patients to make trade-offs between outcomes.(Elwyn et al., 2010; Karkazis et al., 2010; Siminoff & Step, 2005; Steering Committee, 2005) These conditions perfectly

characterize the value-based decisions parents of children with DSD are faced with early on in their child's clinical care.

The tools used to facilitate preference-sensitive care through informed shared decision-making are known as decision-aids. Their efficacy is widely accepted as studies have found that when patients use decision aids they: a.) improve their knowledge of the options; b.) have more accurate expectations of possible benefits and harms; c.) participate more in decision making; d.) experience more positive patient-practitioner communication; e.) select choices that are more consistent with their informed values (when the decision-aid includes values-clarification exercises); f.) are less likely to choose elective surgery interventions (in cases where elective surgery is an option) after they consider other options; and g.) experience positive effects on long-term outcome measures. (Joosten et al., 2008; D Stacey et al., 2011) If a decision-aid tailored to parents of children with DSD brought about even *one* of these effects listed above, DSD care would be greatly improved.

Today, there are over 500 patient decision-aids available or being developed to assist patients making difficult medical decisions for a variety of conditions. It's time a decision-aid specific to parents making early medical decisions for their children with DSD is added to this repertoire. The decision-aid outline presented in Chapter 4 represents the first step in the development of this DSD-specific decision-aid. It was created in conversation with the larger discussions about informed decision-making and shared decision-making in this clinical context. You may not agree with everything that I said, or everything that I proposed. You may even think that you have a better way to achieve the goals of informed shared decision-making via the use of a decision-aid in this context. I welcome any and all suggestions to improve what has been presented. Like any good

decision-aid, the purpose of this thesis was to foster conversations – to provide a framework and starting points for future discussions.

A collaborative effort is needed to bring this DSD-specific decision-aid outline to fruition. Future development and implementation of this decision-aid will require the support of clinicians, activists, scholars, people with DSD, and the parents that inspired its creation. In parents' words I heard a call to action. To the best of my ability, I have tried to answer their call. But I am only one person, so I leave you with this. A call to action: Use the discussion and the DSD-specific decision-aid outline presented here as a catalyst for future development. Together we can make a decision-aid specifically tailored to the early decisions parents must make, and help ensure that parents are making the *best* decisions for their children with disorders of sex development.

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