WHAT HOSPITALISTS SHOULD KNOW ABOUT INTERSEX ADULTS

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ABSTRACT An intersex condition is one in which an individual is born with atypical male or female external genitalia, gonads, hormones, or chromosomes. Older terms, such as pseudohermaphroditism and hermaphroditism, are controversial, leading the Lawson Wilkins Pediatric Endocrine Society (LWPES) and the European Society for Paediatric Endocrinology (ESPE) to propose in 2006 the expression "disorders of sex development" (DSD), a contentious term in its own right, to define congenital conditions in which development of chromosomal, gonadal, or anatomical sex is atypical. The birth of a child with DSD often prompts a long-term, multidisciplinary strategy, involving an array of health-care professionals. While much has been written about these potentially challenging management decisions, particularly those regarding infant genital surgery, very little has been written on the health management of intersex adults, and no literature exists regarding the role of the hospitalists—specialists in inpatient medicine—in the care of patients with DSD. With more than 50,000 members, Hospital Medicine represents the fast-growing subspecialty in the history of medicine. We believe hospitalists are uniquely positioned to provide high-quality care while educating trainees about issues pertinent to this often marginalized patient population. This essay poses a case study to explore some of the medical issues that intersex patients face as adults and that hospitalists will likely encounter.

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A 35-year-old woman presents to the hospital after a fall at home. A routine medical history and physical examination reveal that the patient identifies as intersex, and an X-ray of the left hip demonstrates profound osteopenia. The patient is admitted to the hospitalist service for further evaluation.

What does it mean to identify as intersex? In the medical world, "intersex" is usually referred to as DSD, or "disorders of sex development." Until the 1990s, physicians referred to this condition as hermaphroditism, but this label is no longer used because it evokes mythical creatures rather than real human beings. Even the DSD label is controversial, and the patient in our case study does not think of herself or her condition as a "disorder," merely that she was born with atypical sex development. For some people, this means that they are born with genitals that look ambiguous enough that physicians and parents have difficulty determining what sex to put on the birth certificate. For others, their intersex condition, in this case androgen insensitivity syndrome (AIS), is discovered as the patient gets older, either in childhood (often appearing as a "hernia" that leads to the discovery of internal testes) or later at puberty.

Hospitalized patients are increasingly cared for by hospitalists, and as we have written elsewhere, we believe it is essential for these clinicians to be conversant in issues of gender and sexual identity, both to enhance the patient-doctor relationship and medical education, and, potentially, to improve outcomes (McCarthy, Reis, and Fins 2016). We believe the same is true for differences of sex development, which are rarely discussed on inpatient rounds during the clinical years of medical school. In fact, most practicing clinicians have received exceedingly little training in the care of patients with DSD. In this article, we use a clinical vignette to explore the ways in which hospitalists might change that, both to improve the quality of inpatient care and medical education.

TAKING A THOROUGH MEDICAL HISTORY

The patient in our vignette has presented to the hospital after a routine fall and was noted to have an unusual clinical finding: osteopenia. While the condition is common in adults over 70, it is unusual in an otherwise healthy 35-year-old woman. The patient is admitted to the academic hospitalist service and will be cared for by a team of health-care workers, which include a third-year medical student, a medical intern, a second-year medical resident, and an attending hospitalist, as well as a group of nurses, social workers, and physical therapists. The hospitalist introduces the team to the patient in the emergency room and begins the encounter as she would with any other patient by performing a complete medical history.

¹Some people prefer to explain DSD as "difference of sex development." For an analysis of the complexity of naming intersex, see Davis 2015 and Reis 2007.

Our patient was born with typically female appearing genitals, and so there was no question about her sex. It wasn't until she reached puberty and never started to menstruate that her parents took her to their pediatrician. An ultrasound revealed that instead of ovaries and a uterus, our patient had internal testes. Further testing revealed that she had XY chromosomes, rather than XX chromosomes typically found in females.

As a 15-year-old, this young woman presented characteristically of patients who have complete androgen insensitivity syndrome (CAIS). Patients with CAIS do not respond to androgens at all, so despite their XY chromosomes, their bodies develop along a female pathway. Neither our patient nor her parents would have had any reason to suspect she had AIS unless there were other women in the family with the condition, as AIS is a sex-linked recessive trait affecting the androgen receptor (AR) gene that can run in families.

The hospitalist makes note of this and follows her careful medical history with a review of the woman's surgical history. Twenty years ago, our patient's pediatrician had recommended that she undergo surgery to remove her internal testes in order to prevent them from becoming cancerous. The hospitalist remarks to her team that this is no longer the standard treatment, because physicians now believe that the cancer risk is very low (1% in childhood and slightly higher after puberty, 5–10%; DSD Families 2016). Though girls with CAIS do not respond to the testosterone that their gonads produce, the testosterone is transformed into estrogen, which stimulates breast development and supports bone health. For these reasons, patients today are increasingly given the option to keep their gonads; those who want them removed have to take hormone pills to maintain their bone health.

But our patient was born in 1981 and diagnosed in 1996, when gonadal removal was still common for all such conditions. No one told her or her parents exactly what was going on, however. Not wanting to alarm her, the pediatrician told her euphemistically that her body "had not become fully female," and he never revealed the exact nature of the surgery. Both the surgery itself and the secrecy were common practice in the second half of the 20th century, but both have come under scrutiny in the last 20 years. This information is crucial for understanding why the patient may have developed osteopenia at such a young age.

The patient appealed to her parents for more detailed explanation, but they too did not fully understand the pros and cons of removing the testes and only wanted to ensure that their daughter wouldn't get cancer and die. In retrospect, our patient thinks that her parents may also have wanted to get rid of any trace of "maleness" that those testes represented in their daughter's body, and so they consented to the surgery. Due to the recent efforts of intersex activists, today's protocols have improved, and psychological counseling would be recommended for patients and their families as they contemplate decisions around surgery.

Because the patient no longer had testes post-surgery, she no longer produced any natural hormones on her own and would need to take synthetic hormones

for the rest of her life. She was prescribed estrogen at age 15 and took it for three years until she went to college. While at college, she went to the campus health center to get a new prescription and encountered an uninformed and insensitive physician. The doctor insisted on giving her a pregnancy test, even though she told him that it was not necessary, as she had no ovaries or uterus. In addition, the physician was unfamiliar with AIS and showed no interest in learning about it. Instead he asked her invasive and embarrassing questions about her genitals and her sex life. Our patient left in tears, hasn't been to a doctor since, and she hasn't taken any hormones at all for the last 20 years.

The attending hospitalist mentions to her medical team that this scenario typifies how physicians have long handled cases of intersex. Beginning in the 1990s, intersex activists have drawn attention to the problematic medical practices that have characterized this field, but unfortunately they have been unable to alter this script completely, and many intersex adults still encounter insensitivity and unfamiliarity. Much of what we hear today about intersex concerns the treatment of infants and toddlers. Because atypical genitals at birth are obvious, and physicians have been trained to surgically "fix" them, they have sought to remove ambiguity and to clearly distinguish male from female, surgically "correcting" bodies that did not conform to "normal" male and female anatomies. Since the mid-20th century, such infants routinely underwent "normalizing" surgery, which in fact did not erase the intersex conditions and instead caused lasting physical and psychological harm, including the loss of sexual and reproductive function, as many intersex people have attested (Foundation for Narrative Inquiry 2016).

The hospitalist informs her team that patients born with AIS may have vaginal hypoplasia, which means that the vagina is not deep enough to accommodate heterosexual penetration. This can be discovered at any time in a girl's life (typically if the testes create a hernia, which necessitates surgery), and in the past the response was to surgically create one through vaginoplasty, sometimes in infancy. Parents of young children, in particular, have complained about the post-operative care, as the newly created vagina has to be dilated three times a week for years after the surgery, until (and if) the patient is heterosexually active—a painful process that both young girls and their parents have found unbearable. As the only reason to create a deeper vagina is for sexual penetration, intersex activists have urged physicians to wait until their patients were older, so that they could make their own decisions about their bodies and their sex lives. Since the 19th century, physicians have wanted to surgically "correct" genitals to match deep-seated notions of normal bodies, often elevating marriage as a goal to parents considering surgery for their children. Heidi Walcutt (1999), in an autobiographical essay, recalled that

²As an alternative to vaginoplasty, teenage and adult women may prefer vaginal dilation. See, for example, Bach 2011.

³For critiques of how surgical "correction" has caused more harm than good, including further surgery to eliminate scar tissue and procedures that resulted in the loss of sexual sensation, see Devore 1999 and Nicholson 1999.

in the 1970s a doctor at the Buffalo Children's Hospital told her at age 14 or 15 that she would need surgery to increase the depth of her vagina "if you ever want to have normal sex with your husband."

Intersex activists have spent the last 20 years trying to convince doctors to stop performing corrective surgery, to wait until the child can have a say in operative procedures. They recommend choosing a sex based on hormonal and chromosomal factors in cases of ambiguous genitalia, but delaying any surgeries other than life-saving treatments. An intersex birth is not generally a medical emergency, but it has been seen as a social emergency necessitating clear division between male and female. As the gender scholar Suzanne J. Kessler (1998) has declared: "Gender ambiguity is 'corrected,' not because it is threatening to the infant's life, but because it is threatening to the infant's culture" (32). This unfortunate history of nonconsensual surgery has been destructive in many ways for intersex patients and their families.⁴

In our patient's case, her intersex traits were not known until she was a teenager. Though she did not face vaginoplasty, the moment she found out that she had XY chromosomes and internal testes marked the end of her trust in the medical profession. Her doctor had never told her the truth about her body and what to expect as she grew older. Even the threat of cancer may have been more exaggerated than real; internal testes can be monitored, and today many physicians recognize that they do not need to be removed for fear of cancer—particularly in cases of CAIS.

Our patient's current condition of severe osteopenia can be partly attributed to the unfortunate lack of quality health care for intersex people. If the college health center physician had known more about AIS, he might have explained the importance of estrogen to our patient, and his inexcusable behavior would not have turned her against the entire medical system. She felt like a freak in that setting and has not been eager to repeat the experience, even though she knows that estrogen is important for bone health.

MEDICAL MANAGEMENT AND FOLLOW-UP

Fortunately, the hospitalist was familiar with intersex/DSD and explained to the patient that there are several medications that may help restore her bone health, including hormone replacements, vitamin D supplements, and a bisphosphonate. Once the patient is ready for discharge from the hospital, she will be given a follow-up appointment with an endocrinologist as well as a primary care physician. The hospitalist also talked to our patient about various reproductive options, as the woman revealed that she is considering starting a family with her husband. Without ovaries or a uterus, she would like to find an egg donor and surrogate, but her suspicion toward the medical profession has lingered, and so she has not

⁴For a history of intersex management, see Reis 2009.

yet approached a fertility clinic, fearing professional inexperience or ignorance at best, and derision at worst, regarding her intersex trait.

CONCLUSION

The needs of intersex patients should become a more integral part of medical school curricula. Estimates put the incidence of intersex at about 1 in 2,000 births, and some people say it is even more common than that.⁵ Not every intersex condition presents itself at birth, as in our patient's case. But even those patients who know about their medical history from an early age have often felt abandoned by physicians who have counseled them and their parents to keep their diagnosis a secret. Secrecy often leads to shame, and that, along with medical decisions made without full understanding and disclosure, can lead to psychological trauma, which compounds the physical suffering that these patients may have endured (Karkazis 2008).

Whether their condition is discovered at birth, as older children, or as adults, intersex patients should be treated with sensitivity and care, like any other patients. Those who have felt unsafe around physicians for years could be risking their health, as in the case of our patient. Fortunately, the patient in our vignette is young. If offered quality care, she has the potential to regain much of her bone health.

Perhaps the current attention paid to the health-care needs of transgender patients will spill over into improvements in intersex medical care. Transgender and intersex are not the same phenomena, but both transgender and intersex patients share some basic needs regarding knowledge and sensitivity on the part of medical providers. At a minimum, both groups of patients deserve autonomy over medical decisions concerning their bodies. For example, in the past physicians have been reluctant to perform chest surgery to remove the breasts of a person transitioning from female to male, if that individual did not also want to take testosterone. The legal scholar Dean Spade (2006) has written compellingly on this subject, describing his own difficulty convincing his physician that he was "really" transgender, despite the fact that he did not want to take hormones, as most transgender men do. This attitude echoes a comparable hesitancy years ago, when surgeons proved reluctant to perform male-to-female surgeries if the transgender women were not able to convince them that they would be "successful" women, which generally entailed an ability to look conventionally feminine and to be heterosexual (Meyerowitz 2006). This kind of medical gatekeeping has been rightly criticized in the transgender community and for the most part is no longer common.

⁵Estimates of intersex incidence vary because there is not widespread agreement about which conditions "count" as intersex, and also because figures have not been gathered systematically worldwide. A study at Brown University projected, based on the compiled frequency of individual intersex conditions, that 1.7% of all births (34/2,000) will be intersex (see Fausto-Sterling 2000).

Intersex people face similar coercion when navigating the medical system. As we have seen in the discussion of vaginoplasty, some women face pressure to accommodate heterosexual sex. Similarly, men with various intersex traits feel compelled to undergo surgery in order for them to be able to stand while urinating or to penetrate their sexual partners. All of these surgeries are social rather than medical. They should not be imposed without the knowledge and fully informed consent of the patients, and it is important for clinicians to understand the context in which they occur. This is also true for intersex infants and children: irreversible medically unnecessary surgery should not be performed without their consent. For both intersex and transgender patients, the cultural norms of heterosexuality and the often oppressive norm of bodily "normality" should not drive medical decision-making. Instead, the patients should be confident that they are receiving all the data necessary to make their own fully informed decisions about how their own bodies look and function, as well as information about the possible psychological outcomes of any recommended treatment.

While there is much work to be done to improve the case of intersex patients, and perhaps some disagreement over how best to achieve these goals, we do believe there are several steps that hospitalists can take now. First, intersex/DSD should be incorporated into teaching sessions on inpatient clinical rotations at academic medical centers. Rather than waiting for an intersex patient to arrive at the hospital, academic physicians should help trainees generate differential diagnoses that incorporate the spectrum of sex development, even in cases that may not appear to be related to DSD. This will help foster an environment of acceptance, further academic inquiry, and help students think broadly about the possible causes of a seemingly routine admitting diagnosis.

Second, hospitalists should become active in medical school curriculum development. Providing clinical context and perspective is invaluable to students who are learning about intersex for the first time. In addition, hospitalists should make physicians and medical students aware of the negative effect of using stigmatizing language such as "disorder" of sex development. While some patients are comfortable with the term "DSD," there is a growing majority who are not, and use of such language can be counterproductive to the doctor-patient relationship. Hospitalists can also suggest psychological care for intersex patients and parents, including referral to the many support groups that exist today. Lastly, hospital medicine departments should invite DSD experts, including intersex individuals, to give formal or informal lectures, which may include medical grand rounds and house staff morning reports, as inter-department case conferences. Fostering an inclusive environment will inevitably lead to an exchange of ideas that will benefit all who participate in the delivery of medical care.

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