

## HEALTH BEAT

LOUISE FLEMING

# Which Way to Go?

**B**EING A PARENT is not easy. Parents are confronted every day with difficult choices they have to make for their children. Do I need to take my daughter to the doctor for her fever? Is hockey or soccer a good sport to play even though there is a risk of my son getting hurt? Should I let him drive at night despite the approaching storm? These are all decisions that parents make, often after seeking information from their families, friends, and health-care providers. They weigh the pros and cons, listen to advice offered, make decisions, and hope for the best. However, there are certain decisions some parents must make that they never imagined they would be confronted with. Friends cannot really offer advice, and doctors only can present them with imperfect options, which they know will greatly affect their child—one way or another—for the rest of his or her life.

When a girl is born with classical congenital adrenal hyperplasia (CAH), parents often must make this type of decision—the kind that keeps them up at night; that they cannot post on Facebook to see what other parents did when faced with the same situation. The choice involves whether or not their daughter should have surgery to correct the atypical genitalia she was born with.

CAH is an inherited endocrine condition that affects males and females and places those affected at risk for adrenal crisis. There is no cure, although daily treatment with certain medications is effective in controlling the condition. Babies born with CAH have elevated levels of testosterone throughout gestation that decrease to normal levels after treatment has begun. Some girls with CAH are born with atypical genitalia due to the elevated testosterone exposure during gestation. Despite this anomaly, these girls are XX individuals with functioning ovaries, fallopian tubes, and internal anatomy, with the capability of conceiving. The vast majority of these girls develop in adolescence similarly to their non-CAH-affected peers and identify as females throughout their lives.

CAH also is a life-threatening condition for all who are affected. They must be given additional medication or an intramuscular injection when acutely ill to prevent a potentially fatal adrenal crisis. After parents of a newborn baby girl with CAH have digested this frightening aspect of the condition, they must process how best to respond to their daughter's atypical genitalia.

If they hold off surgery (which may include surgical reduction of the clitoris and construction of a vagina and labia) until their daughter is old enough to make the decision for herself, there is the unknown effect of her growing up with genitalia that are not congruent with her gender identity. This has the potential to create a lifelong body image disturbance. They may question if their daughter can still wear a bathing suit or ballet leotard without feeling stigmatized. Can she still change in front of her friends in the locker room or when trying on clothes in a store's dressing room without feeling shame and embarrassment be-

cause her genitalia look different? Will she be bullied by her peers?

On the other hand, if they opt for the surgery when their daughter is young, will she resent that decision later as she grows into adulthood? Will she have altered sexual satisfaction as an adult because of the surgery? Will she require repeated surgeries as she enters adolescence—and do they even have a choice in avoiding surgery at all? For some girls, early surgery may be medically necessary to prevent irreversible damage from urine backing up into the bladder and kidneys.

These are not easy decisions for any parent to make. However, when CAH is misrepresented in the public arena, as it has been over the last several months, it makes a complicated and challenging decision even more traumatizing for families facing this dilemma. CAH has been portrayed as an intersex condition, which the CAH community has repeatedly rejected. It also is not the only condition that causes a baby to be born with atypical genitalia. In addition, girls born with CAH do not experience atypical genitalia uniformly, but rather with varying degrees. For some, surgery would not even be considered by the parents or medical community.

There are women with CAH who are very satisfied with the outcome of their surgery and grateful that their parents made the decision early on

in their lives. There also are parents of young girls who made this difficult decision in the last five to 10 years who feel confident that their daughter received expert surgical care and are secure with their decision, which was made in an informed manner with experienced health-care providers. The narrative that has been presented to the public (as in the documentary, "Gender Revolution: A Journey with Katie Couric" that aired on the National Geographic Channel)—whether it is presenting CAH as intersex, vilifying the parents who opt for early surgery or the surgeons who perform it, or ignor-

ing the life-threatening nature of the condition—must stop now.

CARES Foundation, an organization that leads the effort to improve the lives of the congenital adrenal hyperplasia community, wholeheartedly supports a thorough discussion of CAH. CARES encourages and assists with research endeavors that explore the surgical outcomes—both positive and negative—associated with atypical genitalia. CARES also supports all groups of people that feel misrepresented and marginalized, including the LGBTQ community. Finally, CARES recognizes that surgical decisions for girls born with CAH are intensely personal, inherently complex, and, at times, deeply divisive. However, any discussion of CAH should be medically accurate and fair, without bias or agenda—delivered by specialists in the field.

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