Published in final edited form as:

J Med Humanit. 2012 September ; 33(3): 209–212. doi:10.1007/s10912-012-9176-6.

Healing and Harming

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Abstract

As part of a volunteering program, a medical student learns the story of a sick infant whose survival was wholly dependent on modern medicine. The family was pacified by believing, during pregnancy, that the fetus would be healthy. Poor counseling and unethical physician behavior led to the trauma of the family's struggle to keep its baby alive, happy, and as healthy as possible. The need for adequate, thorough, and individualized counseling is a critical and still unfulfilled need in patient care.

Keywords

urethral obstruction; fetus; ethics; kidney failure; transplantation; perinatology

In 2003, Sarah was happily pregnant with her second child and enthusiastically working full-time as director of a foster family agency. Fifteen weeks into the pregnancy, an ultrasound revealed that the fetus' bladder was distended to fifteen millimeters. Sarah's obstetrician told her there might be a problem with one of the kidneys but reassured Sarah that the baby only needed one healthy kidney and that the normal amniotic fluid level indicated that the current kidney function was adequate. He recommended that Sarah consult with a perinatologist.

Sarah and Dave consulted a perinatologist who Sarah believes manages "every high-risk pregnancy of the celebrities." With Sarah and Dave, he was abrupt and seemed distracted. He reiterated what the obstetrician had said: that one kidney appeared to be at risk, but the other looked healthy, and the baby should be fine. The specialist also advised them to see a pediatric urologist.

The pediatric urologist told Sarah and Dave that one in two hundred babies has the same problem, and theirs would be healthy after a simple surgery at birth. He failed to explain that, though he may have been quoting statistics for hydronephrosis, which is common, Sarah and Dave's fetus already had a diagnosis of probable posterior urethral valves, which has an incidence of one in 1,250 fetal ultrasound screenings and one in eight thousand to one in twenty-five thousand live births (Casale 2007).

Sarah and Dave, concerned about the well-being of their unborn child, were comforted by the reassurance of a healthy baby by all three specialists they had consulted. They did try to discuss with their doctors the possibility of terminating the pregnancy, but none of the doctors – the obstetrician, the perinatologist and the urologist – recommended or offered termination as one of the medical options. Sarah and Dave opted to continue the pregnancy, and Sarah underwent routine prenatal healthcare along with bimonthly ultrasound evaluations.

At thirty-one weeks of the pregnancy, an ultrasound revealed severe oligohydramnios, and Sarah was immediately hospitalized for possible urgent delivery and corticosteroid treatment. Adam was born two days later by cesarean section and immediately admitted to the Neonatal Intensive Care Unit (NICU). When he was one day old, an ultrasound revealed for the first time that in addition to Adam's dilated ureters and bladder, he had bilateral dysplastic kidneys. The planned laser valve ablation was performed successfully, but Adam's problems were far from over. He became hypotensive and was placed on respiratory ventilation, in end-stage renal failure.

Terrified, Sarah and Dave spent most of their time by Adam's bedside. They were unable to hold their infant until he was two weeks old. The nephrologist, neonatologist, and other team members finally met with the parents to give them the dire prognosis. Dave remembers hearing that Adam's kidneys were in complete failure, and he was unconscious because his blood was "poisoning his brain." The team of doctors recommended that Adam start dialysis, but for the first time seemed to encourage Sarah and Dave to decline this recommendation.

Dave relates, with tears in his eyes, "There's only one choice a parent can make." After touching and holding his little boy, Dave felt an unshakeable bond and urgency to do everything to help Adam. Sarah and Dave elected to proceed with the dialysis in order to give Adam his best chance at ultimately receiving kidney transplantation and potentially a full life. Adam was maintained on peritoneal dialysis and eventually discharged home after two months of hospitalization.

Sarah and Dave are certain that the specialists they consulted during pregnancy had never warned that posterior urethral valves is a dynamic condition for which the probability of renal failure is significant but cannot be determined prenatally with accuracy. If mentioned at all, kidney transplant was not a prominent feature of the prenatal urology consult. They felt blindsided by the severity of Adam's illness.

At home, Adam required intensive care, including peritoneal dialysis. With no medical background or the resources for a dialysis nurse, Sarah handled her son's nursing with meticulous care. She quit her job and put her career on hold. Dave remembers more poignantly the acute and life-threatening emergencies. Adam frequented the emergency room and pediatric wards of the hospital due to recurrent infections and surgical complications. Dave also recalls Adam's fever-induced seizures and even a post-operative code during which Adam nearly died in his arms.

When Adam was just over two years old, Sarah and Dave received a late-night call that Adam would finally get a kidney transplant. He survived the surgery but sustained multiple complications. After a prolonged recuperation, the new kidney eventually responded and began to function on its own, but this solved just one of Adam's problems. His significant developmental delays required physical, occupational, social, and behavioral therapy. He continues to have chronic medical issues that need constant care, including regular maintenance of a gastrostomy tube from which he has never been able to wean. The

prospect of significant lifelong developmental delay from kidney failure and dialysis at a young age was never discussed in the many prenatal consults and routine exams.

Adam would not have survived without multiple sophisticated medical advances. First, high-resolution ultrasound was necessary to diagnose and monitor his condition. The posterior urethral valve ablation was performed immediately after birth to correct the anatomical birth defect. Neonatal intensive care, dialysis, and allogeneic renal transplantation further prolonged his life. Today, seven years after Adam's birth, all these interventions are a testament to modern medical science. Though he is now medically stable, he is incontinent, relies on tube feeding and will need immunosuppressive therapy for the rest of his life. He will always have an intellectual disability, and he requires a full-time aide and a special school. Without the social ability to participate in cooperative play even with his brother, Adam has no friends. His mother has never returned to work because of the demands of caring for him.

Posterior urethral valves is the most common condition in the broader category of obstructive uropathy, which is the largest single cause of renal failure requiring transplantation (Peters and Chevalier 2007). Renal failure develops in seventeen to seventy percent of children with prenatally diagnosed posterior urethral valves, and death or renal failure in forty-five to fifty-three percent of those diagnosed before twenty-four weeks of gestation (Hutton et al. 1994; Roth, Carter and Chan 2001; Sarhan 2008). The introduction to the chapter, "Posterior Urethral Valves and Other Urethral Anomalies" in *Campbell-Walsh Urology* states, "Congenital obstruction of the urethra is one of the most devastating anomalies to occur in the urinary tract and one of the few that are life-threatening in the neonatal period. These lesions usually result in lifelong disabilities with incontinence and decreased renal function despite optimal medical management" (Casale 2007). In a newly-published series of sixty-five cases of posterior urethral valves, all were diagnosed after twenty weeks of gestation, so Sarah's diagnosis at just fifteen weeks was unusual and ominous, signifying a fifty percent risk of renal failure (Sarhan et al. 2008; Hutton et al. 1994).

Why did the doctors reassure Sarah and Dave to expect a healthy baby? Why were Sarah and Dave not informed and educated about possible complications and poor outcomes? Why were renal failure, kidney transplant and developmental delay hardly mentioned, if at all? Would Dave and Sarah have made the same choices if informed properly and prepared adequately for the worst case scenario? Would they have chosen to bear a child who had to endure such extraordinary pain and suffering and then live with permanent disabilities?

Clinical practice guidelines have been shown to improve patient care and outcomes, but few general patient counseling guidelines have been developed for parents during the prenatal period (Grimshaw and Russell 1993; Lugtenberg, Burgers and Westert 2009). A 2001 medical review of posterior urethral valves advised that "decisions [regarding termination of pregnancy] should be made after counseling with regard to realistic long-term expectations of renal and pulmonary function" (Gatti and Kirsch 2001). However, professional guidelines on patient counseling from the American Medical Association and the American College of Obstetrics and Gynecology address only specific high-profile issues such as prenatal genetic counseling and decisions about life-saving care of newborns (AMA Opinion 2.12 2009; AMA Opinion 2.215 2009). These or other authoritative bodies should establish practical and ethical guidelines for patient counseling to encourage appropriate care.

Dazzling technological advances in medicine allowed Adam to survive because of a prenatal diagnosis and miraculous medical and surgical interventions. With his parents deprived of

the opportunity to make an informed decision about the fate of their son who now faces a lifetime of disability, the case illustrates the consequences of inadequate patient counseling.ⁱ

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ⁱSarah and Adam's physicians were not interviewed for this article. At least one perinatologist, pediatric urologist, and pediatric nephrologist, none of whom were involved in Adam's care, were consulted about this essay for medical information and views. Sarah and Dave have read the article and given their permission for publication. However, to protect their privacy, all names have been changed.