

**Through the Land of Oz:**  
*Self-Advocacy in Today's Health Care System*  
by  
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In January 2003, the cardiologist declared, “You should have your babies right away. Your heart valve will need replacement and it will be difficult, maybe impossible, to have kids after your operation. You have the choice of a pig valve or a mechanical valve.” My daughter, Hilary Engelman, was 23 years old, not married, and not yet thinking of having children. After the cardiologist’s pronouncement, we cried, not for the only time on this medical odyssey.

Hilary had been diagnosed at age seven with mitral valve prolapse (MVP). Since she had no symptoms, doctors relied on echocardiograms - sound waves that create a moving picture of the heart – to give an accurate, quick assessment of her overall heart function. Initially, they thought her condition was a benign heart murmur, the case for many people with prolapsed mitral valves. Three years after first diagnosis, though, a pediatric cardiologist at our health maintenance organization (HMO) said, “Your daughter’s MVP is not trivial and may require surgery someday.” With a hint of sadness in his voice, he pointed to a photograph of a vibrant-looking teenager on his bulletin board and added, “A patient of mine died from a leaky valve like hers.”

I had been a member of this HMO since before Hilary’s birth. Both my children were born there and we had received great care. I trusted them to follow her condition vigilantly, but this physician’s deadly comparison irritated me. It also got my attention and started us on our long journey toward understanding the exact nature of her heart valve problem and how to fix it. I did what any mother, any consumer can do – I stubbornly sought every piece of information

available on MVP. For three days following that appointment I read and cried, cried and read, relieved to uncover facts but fearful for the future.

I found that the medical phrase “mitral valve prolapse” referred to a condition more than a diagnosis, describing a range of heart valve variation, from benign to malignant. The mitral valve controls blood flow between the upper and lower chambers in the left side of the heart. Blood should flow only one direction, from the upper chamber into the lower. If these valve flaps do not work properly, part of the valve may “balloon” into the upper chamber and regurgitate, or leak.

Over the years, doctors applied a dozen descriptors to Hilary’s malfunctioning heart valve, including deformed, flailing, floppy, and, her least favorite, ugly. For consumers like us, this list did not cast more light on her condition. We knew only that her valve was “prolapsed,” and a prolapsing valve does not necessarily mean anything medically important. Many people walk around with prolapsed mitral valves and never need treatment, much less open-heart surgery.

Cardiologists, who treat disorders of the heart that can be managed non-surgically, followed my daughter’s case over the years. With no symptoms and semiannual passable echocardiograms, we slipped into a shared, dangerous illusion of good health. But with the 2003 warning that Hilary should have her babies right away, surgery suddenly hung over us again. I arranged for several surgical opinions, and during this round of consultations, a cardiac surgeon first uttered the dreaded words, “Your daughter’s valve is probably not repairable.” But at the time, cardiologists and cardiovascular surgeons said she did not need an operation, at least not yet.

Then, in December 2007, two HMO cardiologists warned that surgery should happen within a month. Although we had long expected this day, its arrival overwhelmed us. Hilary said to one doctor, “You have no idea how hard it is agreeing to surgery when I have no symptoms. I feel fine!”

Compared with her echocardiogram from six months earlier, her valve now leaked severely, creating two jets - an ominous finding. Blood pressure increased in the arteries supplying her lungs, and her heart muscle showed signs of exhaustion. If leakage forces the heart to work too hard to pump blood, its function will eventually decline. Damage to a heart muscle is irreparable and permanent. Her primary cardiologist warned that shortness of breath, dizziness, and fainting on exertion were not far away and immediately referred her to our HMO’s cardiovascular surgery department to schedule a heart operation.

Her heart murmur had turned out to be more than a variant of normal. It was on the severe end of the MVP spectrum, and, uncorrected, a valve like hers may lead to a heart transplant or even death. Hard enough to think she would require open-heart surgery, but knowing she had the kind of valve that her doctors considered too difficult to repair left us numb.

Clinging to the life raft of current literature, most of it available on the Internet, I discovered critical information. Her defective valve might actually have a name: “Barlow’s valve,” a consequence of “Barlow’s disease,” first written of in the 1960s. None of the HMO physicians had mentioned this label, though it fit uncomfortably well with their many descriptions of her troubled valve. I was relieved to find a possible diagnosis, hoping that a specific label for her valve might also mean a specific treatment.

Second, I found that the likelihood of successful childbirth lowers after mitral valve replacement. When I initially understood that my daughter’s heart condition might require

surgery, a friend, a cardiac-nurse specialist, had mentioned that mitral valve repair, not replacement, was the best surgical treatment. She said that too many patients with severely-leaking heart valves receive replacement valves instead of repairs. With mitral valve repair superior to replacement, the choice between the two matters to all patients and is a crucial detail for a woman, such as Hilary, who is yet to have her children.

Third, and perhaps most significantly, I discovered why physicians had not given Hilary's prolapsed mitral valve a more specific label. The reason stemmed from uncertainty in classifying valve problems. Even among cardiologists and cardiac surgeons this confusion can keep doctors from understanding each other and patients from knowing how to choose a qualified surgeon. Hilary's specific difficulty would identify the type and complexity of lesions that her surgeon would likely encounter in the operating room and the level of surgical skill needed. And, critically, choice of surgeon would largely determine whether her valve was repaired or replaced. But so far, cardiologists and cardiac surgeons had focused solely on the state of her valve, not identifying choice of surgeon as a key issue in whether her valve would be repaired.

I also found expert opinions supporting mitral valve repair over mitral valve replacement. Dr. Bernadine Healy is a cardiologist and former head of the National Institutes of Health and the American Red Cross. She discussed severely leaking mitral valves like Hilary's in *US News and World Report*, October 8, 2007. Paraphrasing, this article said that patients with repaired valves live longer, with better heart function and less risk of complications; that in the right hands, more than 90 percent of leaky valves can be repaired, though those in the U.S. actually getting repairs is lower, closer to 50 percent; and that most cardiac surgeons do only a few mitral operations each year, primarily replacements, mainly because replacements are what they know

how to do well. Dr. Healy also said that repair surgeries require extraordinary knowledge and judgment; that tougher cases of mitral valve disease require the skill found in high-volume heart centers; and that if done properly, the fixed mitral valve functions almost as well as a normal one.

In 2006, the American College of Cardiology/American Heart Association (ACC/AHA) had published their Guidelines for Management of Patients with Valvular Heart Disease; they recommended MV repair over replacement for the majority of patients requiring surgery for severe, chronic valve leakage. The same year, surgeons in England drew the same conclusions (Ray et al, 2006).

With this encouraging information in hand, we focused on finding a surgeon who was skilled at repairing a severely deformed valve like Hilary's and who could also advise us about timing her surgery. According to the British surgeons, to improve a person's health, valve repair must eliminate valve leakage before onset of symptoms leading to heart failure. If a patient waits too long, even "normal" echocardiograms can mask significant heart muscle dysfunction and onset of symptoms may go unnoticed. Once a heart begins to fail, even if a successful repair follows, the patient's life span is likely shortened. And when an operation occurs either too late in the course of the disease or as an emergency, repair is less an option and replacement more likely. Timing my daughter's surgery required walking a very fine line.

Following our meeting with the cardiologists who told Hilary she needed surgery right away, HMO staff had stressed that the cardiovascular surgery department was "remarkably sophisticated." But our choice of surgeons narrowed dramatically when we learned that only a few of them actually operated on heart valves. We selected the only plan surgeon who, by reputation, was skilled in mitral valve repair, with more experience at valve surgery than the

others. His staff told us that patients came from all over the country for him to operate. With three long weeks until our appointment with this surgeon, and anxiety a permanent companion, I moved things forward from another direction.

I sent copies of two of my daughter's most recent echocardiograms to a cardiac surgeon in New York City. Dr. David Adams is affiliated with a specialty, high-volume heart center such as Dr. Healy mentioned in her article, the Mitral Valve Repair Reference Center at The Mount Sinai Hospital. I chose Dr. Adams because I appreciated the quality of his writings on mitral valve repair. I asked if Hilary had Barlow's disease, whether he thought she needed surgery, and if so, when? Most important, I asked if he considered her valve repairable. He responded quickly: Yes, it was a Barlow's valve, she needed surgery, and, yes, in his opinion her valve could be repaired. He also said: "This is an operation for 2008, not 2009."

I felt agitated that surgery seemed truly upon us, but calmed to know my daughter's valve might be repaired. I admit to feeling a bit hesitant about his response, however, since Dr. Adams' assessment countered numerous other physicians who had told us that her valve would likely have to be replaced, not repaired.

When we entered the most-experienced HMO cardiac surgeon's office in January 2008, I looked forward to his estimate of best timing for surgery and fervently hoped to hear him say that she had a repairable valve. We had been told by experts that Hilary's was not garden-variety mitral valve prolapse, but would this surgeon confirm Barlow's disease? I assumed he would not only be familiar with Hilary's records but also sensitive to a young woman facing open-heart surgery.

Wrong on all counts. He appeared not to have reviewed my daughter's medical records. He fumbled through chart notes and could not make his computer work to study her

echocardiograms. We sat expectantly. He gave her records a cursory look, then said she did not need surgery and should schedule another echocardiogram in six months. We stared at him. His statement contrasted sharply with other experts, many within his own organization who said she needed surgery now. He then said she had no symptoms and grimly added that surgery had many dangers and she “might have a stroke or die” during or as a result of surgery.

He reviewed a series of catastrophic possible outcomes in extensive, insensitive detail - failed surgery, death, loss of biologic function from stroke, heart or kidney failure, additional surgeries with unexpected outcomes, and on and on. We had lived with the fear of those possibilities for many years, but he spoke as if he was giving us new information. No one in her right mind would voluntarily undergo mitral valve surgery after his litany. Yet, in the face of these possibilities, and all too soon, Hilary would have to muster the emotional courage to tackle what only she could face: open-heart surgery.

Why did he focus on the risks of surgery, not the benefits? Delaying surgery, too, carried potentially grave hazards, but he said nothing about them. All the HMO cardiologists seemed concerned about my daughter waiting too long, but this surgeon did not. Did these people ever talk to each other? I felt stranded in the Land of Oz, baffled by the smoke and mirrors of the Wizard, invited to trust the man “behind the curtain” and give up what I knew to be true. I felt his opinion begin to erode my own judgment, based on knowledge from several sources, but forced myself to speak. I asked about “Barlow’s valve.” At first, he did not acknowledge the question, but a few minutes later, said off-handedly: “It might be a Barlow’s valve.”

Determined, I forged ahead with other questions prepared in advance - the what, when, why, and how. Surgery - when and why and how long does it take? Valve repair – what chance in Hilary’s case? Repair versus replacement – how many of each did he do in a year; what was

his repair rate for Barlow's valve? All important information to ask of any valve surgeon and essential to understanding what lay ahead. Certain responses would most likely lead to a timely and successful valve repair, others to delay and a replaced valve.

The surgeon's answers all pointed toward delay. "If and when" Hilary needed surgery, he said, he was qualified to do it and valve replacement was a possibility - her valve defect was "complex and maybe too difficult to repair." He spent time discussing the "several choices" we had for replacement valves. He then said she had only a 60% chance of successful repair, at best, and that he operated on two to three Barlow's valves per year.

We knew that expert mitral valve repair surgeons at specialty centers repaired valves like Hilary's two or three times a week, not two or three times a year. And their success rates were close to 100%, far above his predicted 60%. In addition, the surgeon's focus on her lack of symptoms was a serious distortion, since lack of symptoms does not necessarily mean that a person's heart is healthy. Even when a patient has no symptoms, the 2006 ACC/AHA Guidelines recommend mitral valve surgery in the following case: when her heart still functions well-enough, but she also has chronic, severe mitral valve leakage and elevated blood pressure in the arteries supplying her lungs. Since Hilary met all the requirements of these Guidelines, I had assumed that referral to a mitral valve repair center would be a mere formality, once the surgeon reviewed her records.

But as a result of his decision that she did not need surgery, the HMO would not approve referral. His short-sighted perspective checkmated our ability to seek outside surgical help and, perhaps, was not coincidental. We discovered later that he not only knew about mitral valve specialty centers, but had visited one recently. He chose not to tell us that much better options for Hilary's treatment existed, forgetting or ignoring the part of his Hippocratic Oath that mattered

most at the time: “I will not be ashamed to say ‘I know not,’ nor will I fail to call in my colleagues when the skills of another are needed for a patient’s recovery.”

Why did he give us misleading information? Was he careless in his assessment of her heart condition or simply out of touch with current medical thinking represented by the ACC/AHA Guidelines? Was he not clear about what he did not know? Alternatively, was he clear about what he did not know and, consequently, did not wish to operate on a 28-year-old woman with a severely-distorted valve he knew he could not fix? Was he motivated politically to use “lack of medical necessity” to deny referral and save his organization money? Ultimately, the reason did not matter. His lack of action protected his status and his organization, but put Hilary’s safety and well-being at risk. As a mother determined to protect her child’s health, I could not accept his judgment as final.

We now had two simultaneous challenges: first, to prove to our HMO that she needed surgery as soon as possible and, second, to search for a qualified mitral valve repair specialist who could mend a Barlow’s valve. All this while facing our own emotional reactions to her impending open-heart surgery.

I arranged for additional opinions from surgeons with four major mitral valve repair centers throughout the United States, each surgeon considered an expert at repairing Barlow’s valves. I asked them the same questions I had asked the HMO surgeon. Each confirmed that surgery should happen soon and that repair was likely. Hilary chose the hands she wished to fix her ailing valve.

We requested an out-of-network referral from the HMO’s Medical Center Review Committee. Our request cited supporting medical and surgical opinions, both inside and outside our HMO. It accompanied a labeled, cross-indexed binder, containing our previous

correspondence with the HMO, letters from the consultants that clarified Hilary's deteriorating condition, and copies of literature documenting the need for surgery with a qualified surgical team. The Review Committee, a group of administrative personnel and physicians who are not cardiac surgeons, would decide if open-heart surgery was "medically necessary" and whether to grant referral to an out-of-network facility. The reviewers consulted the cardiovascular surgery department at the HMO, but did not consider the opinions of the HMO cardiologists – the ones who said Hilary needed immediate surgery.

Within days the HMO Review Committee officially and firmly denied our request for out-of-network referral. They said there was "lack of medical necessity" and that their organization had "appropriately privileged and credentialed physicians available to provide this service," if and when necessary.

The Committee referred us to another surgeon inside the HMO's cardiovascular surgery department for a second opinion. No surprise that the "second opinion" was in concert with the first. Surgery wasn't needed, he said, adding that he, too, was qualified to perform valve repair, if required. When I asked specifically about his statistics for Barlow's valve repair, he said irritably, "My statistics are great." I persisted, and eventually he said he did not know for sure what his statistics were, because "if the surgery didn't go well, I wouldn't know because the patient wouldn't return to me, would he?" Presumably the patient died. If this comment was meant to be funny, it failed - we knew that this cardiovascular surgeon had even less experience with mitral valve repair surgery than the first one.

The Chief of Cardiovascular Surgery gave a third opinion. He never met with Hilary, and he agreed with his colleagues.

After three denials that spanned seven months, I felt stumped and again doubted my own hard-won understanding. Had they seen something in the data that I had missed – something that proved Hilary’s situation was not so bad and that she should wait? For my own sanity, I put together a simple chart of her heart measurements, from age seven to the present. The changes were unmistakable, even to a non-M.D. like me.

I contacted San Francisco attorney Arnold Levinson, who had guided our communications with our HMO and, in his unlimited graciousness, not charged one penny for his help. I told him we’d had our final rejection from the HMO, that my newly-created chart of numbers showed we were right about the changes in Hilary’s heart, and I guessed it was time to contact the State Board of Managed Health Care for an Independent Medical Review. “It’s premature,” he said. “Why are you letting the word ‘final’ get to you? If you have new information, your HMO is legally required to consider it.” He then asked about my chart of numbers and whether I could think of one more person in the HMO to meet with – one person to show my numbers, who might be willing to help. I mentioned a retired surgeon. “Go see him,” said Arnie.

We told the surgeon that we had gotten such a range of opinions we didn’t know what to think. Initially, he stonewalled. But as he saw Hilary’s clear distress and, finally, looked at my chart of numbers, he paused.

I said, “We need to know whether this is the time for surgery.” He nodded. “It’s time?” I asked. “Yes, you are in the window when it needs to happen.” Hilary and I both dissolved in tears. After a moment to gather myself, I then asked the second big question: “Should her surgery be done by the HMO?” “No,” he said, “we cannot do that surgery.” He likened the HMO

referring Hilary to a specialty center focusing on Barlow's valves to them referring heart transplant patients to an outside center specializing in transplants.

Within hours of submitting the new information, my chart summarizing years of data and the retired surgeon's opinion, the HMO rejected it, saying, in essence, "What part of 'final' don't you understand?"

This event triggered our last appeal, to the State Board of Managed Health Care. The board referred it immediately to the Center for Health Dispute Resolution (CHDR). Within ten days of receiving my request for an Independent Medical Review, the CHDR over-ruled our HMO's denials. They cited the extensive case file, including all the letters, expert opinions, and research materials that no one in the HMO seemed to have read. The CHDR then said that due to severe Barlow's disease, Hilary needed a complex repair, that only a specialist was qualified to perform it, and that the surgery was medically necessary.

Against all odds, we had succeeded, through stubborn perseverance, self-education, and self-advocacy.

Within days, the HMO contacted Hilary about travel arrangements and lodging while recovering from surgery. Less than a month later, Drs. David Adams and Ani Anyanwu, at the Mount Sinai Medical Center in New York City, successfully repaired Hilary's mitral valve. Her heart valve function is now essentially normal. It no longer leaks, and she has her life back. You might say Hilary found her pair of ruby slippers, in spite of the Wizards' misguided attempts to keep them from her.

Dr. Adams has referred to the increasing opportunity for mitral valve repair as a "revolution." The late mythologist Joseph Campbell said, "Revolution doesn't have to do with smashing something; it has to do with bringing something forth." If mitral valve repair 'brings

forth' the patient's ability to live a healthy life, with her own repaired valve, then they are both correct.

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