

Playing Catch in the Park

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Jonathan was initially brought to an outside hospital emergency department because of a high fever and difficulty breathing. On arrival, he was obtunded and in shock, his blood pressure unobtainable. An intravenous line was placed; he received fluids and pressors, and was transferred to the intensive care unit. An initial portable chest x-ray showed an extensive right lung infiltrate with an empyema later found to contain *Streptococcus pneumoniae*. A central line was placed as well as a brachial arterial line after attempts to place a radial artery line were unsuccessful. The initial laboratory tests suggested hemolysis and renal failure consistent with the diagnosis of hemolytic uremic syndrome (HUS) and the production of neuraminidase, a toxin that destroys the outer protein coat of red blood cells and platelets and leads to kidney damage. In the early 1980s, when this story took place, the fatality rate for neuraminidase-associated HUS was high, well over 50%.

When Jonathan was transferred to our University Hospital Intensive Care Unit to receive peritoneal dialysis, I became his attending pediatrician. When I first met Jonathan's parents Mark and Talia, they were distraught, anxious, and frightened. Mark was a large man with a full black beard, his head covered with a black fedora in the custom of orthodox Jews. He was studying to become a rabbi.

I explained Jonathan's condition and tried to be encouraging but realistic. I told Mark and Talia that Jonathan had stabilized and had made some progress. He'd gotten over the shock, and his infection seemed to be improving. His lungs were also better as shown by his easier breathing. I went on to say that the major challenge now was to try to prevent further damage to his kidneys, and that we hoped the peritoneal dialysis would accomplish that.

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"I think and hope his kidneys will recover," I explained. "But I cannot give you a guarantee."

Mark took a deep breath, and then asked, "Are you saying Jonathan still may die?"

I answered in a low, calm voice. "Yes, he may still die."

I didn't let those words resonate for long before I continued, speaking optimistically about how well I thought Jonathan was doing. I explained that he was stable, but we would closely monitor his blood and platelet counts, blood pressure, urine, and kidney function tests, as well as watch for any signs of congestive heart failure. I admitted that I had only cared for a few children with this condition, so my clinical experience was limited. I closed by saying, "I'm sorry this all sounds so complicated, but it is complicated and I will need your help if we have to make some difficult decisions."

On Sunday, his second day at University Hospital, Jonathan's nurse noticed a problem with his left arm where the arterial line had been inserted at the elbow. The skin below his elbow where the line was placed had become mottled and pale with areas that were black and blue. The arm felt cool and the nurse could not feel his pulse at the wrist. When I saw the purplish mottling of the skin, I knew the arterial line had clotted, obstructing the flow of blood beyond the catheter.

We immediately removed the arterial line. The nurse wrapped the arm in warm towels to try to increase its blood supply. The consulting hematologist recommended against starting heparin, explaining that it would not be effective in dissolving an arterial clot and that it might increase the risk of a stroke. We watched Jonathan closely that night without additional therapy.

The next day there was no improvement. Jonathan's hand was swollen; his fingertips and nails had turned blackish purple. His platelet count had dropped. I asked a hematologist with extensive experience with clotting disorders to consult on the case. He recommended starting a heparin drip but also giving a platelet transfusion. He discussed his recommendations with the parents, explaining that Jonathan might benefit from a trial of heparin. Although the heparin would not dissolve the clot already in the artery, it would prevent formation of additional clots and increase the possibility that new channels of collateral circulation might form. He closed by saying, "It may be a long shot, but it is worth trying."

Remembering the prior conversation, Mark asked, "What about his platelet count and the risk of a stroke?"

The hematologist replied that we would give Jonathan platelets before starting the heparin. Averting the risk of

bleeding from heparin at this point was more important than avoiding further damage to his kidneys from the platelet transfusion.

“What if we don’t do anything?”

The hematologist responded, “If we don’t re-establish some circulation in his arm, we may have to amputate his hand.”

Talia gasped and slowly moved her head from side to side. The thought of Jonathan losing his hand was devastating. Yet the possibility of heparin causing a stroke was equally terrifying. I realized that they needed guidance in making this decision.

I explained that the hematologist was an international expert on the use of heparin in HUS. He had written about it in several pediatric articles. I suggested that they try his approach, recognizing the risks involved.

Mark replied, “Okay, let’s try the heparin.”

Mark then asked if they could hold morning and evening prayer services at Jonathan’s bedside. It was arranged, and later that day Mark, accompanied by several male friends wearing prayer shawls, began chanting a prayer praising God and asking him to cure the sick.

The hematologist counseled patience, saying it would take a few days to see any effect from the heparin. I returned to Jonathan’s bedside frequently over the next 2 days, hoping that the circulation in his arm would improve. Unfortunately it didn’t. The black color in Jonathan’s fingers and thumb was expanding like a stain on a tablecloth. The only other option was to attempt to surgically remove the clot. The cardiovascular surgeon felt that trying to remove the clot would cause more damage, resulting in the loss of all the circulation below the elbow so that Jonathan could lose his entire forearm.

I began to prepare myself for talking to Mark and Talia about possibly needing to amputate Jonathan’s tiny 5-month-old hand. I wondered how difficult life would be for him without his left hand. Both his parents were incredibly strong and loving. I was confident they would help him overcome the challenges of his disability, but it would be a painful and slow process.

Since I was uncertain about what kind of prosthesis and rehabilitation would be needed, I consulted with rehabilitation medicine. The rehabilitation physicians asked the surgeon to reconsider his decision not to operate. They explained there would be little functional difference between losing a hand and losing everything below the elbow. It would be better to try to save the hand than worry about further damaging the lower arm. The cardiovascular surgeons agreed to operate.

The rehabilitation pediatrician and I met with Mark and Talia to discuss the benefits and risks of the surgery. The surgeons believed there was a 30% chance they would be able to remove the clot and restore the circulation, and a 5% to 10% risk of causing additional damage to the circulation so that he would lose his entire arm below the elbow.

“How are you feeling about making this decision?” I asked Mark and Talia.

Mark replied that he wanted Jonathan to have a chance to lead a normal life, playing baseball and other sports with

two normal hands. He wanted to try the surgery and hoped that God would help to make the surgery successful.

The surgeons made an incision at the antecubital fossa, exposing the brachial artery. The artery was totally occluded with a firm clot. Using a special tiny catheter with an inflatable balloon placed over a guide wire, they tried to remove the clot by blowing up the balloon beyond the clot and pulling back. It would not come out. The catheter was inserted further into the artery to try to get beyond the clot and then pull it back. It still wouldn’t come free, so they inserted the catheter further down the artery. Patiently, for the next hour, they gently put pressure on the catheter. Finally they sensed some movement and a spaghetti strand of a clot emerged from the artery. They reinserted the catheter, pulled back, and another strand emerged. After closing the artery, the arm changed color immediately, becoming pinker and less mottled. By the next morning the hand had become pink and almost normal looking, except for the distal parts of his thumb and first two fingers. Those areas were still black and without evidence of any blood supply. He would lose the distal segments of his these digits. Although disfigured, he would still have a functional hand.

Our attention became focused on his kidney function and the need to continue peritoneal dialysis until his kidneys started working. As he started to make small amounts of urine, we reduced the time he received dialysis. One week later we stopped the dialysis. Jonathan’s urine output increased and his blood urea nitrogen and creatinine were stable. He was discharged about a week after stopping dialysis. Few children with such severe neuraminidase-associated HUS had ever survived.

I felt an enormous sense of relief and satisfaction with the outcome of this case as I had grown close to Jonathan and his parents during his hospital stay. In many ways, this was the most challenging case of my career because there had been so much disagreement among the many involved consultants. Several years later, the case took on an unexpected dimension when I was notified that I had been named in a lawsuit.

Mark filed a lawsuit claiming medical malpractice related to the loss of the distal segments of Jonathan’s thumb and fingers. I was stunned and angry. I, along with the cardiologist who had placed the brachial arterial line, and the 2 hospitals where Jonathan received care were named in the suit. The University provided me with a lawyer, who advised me not to have any contact or conversations with Mark or Talia. He explained it was common in malpractice cases to name everyone and every hospital that had cared for the patient in the lawsuit. Mark and Talia’s attorney submitted the names of the expert witnesses who would review the medical records and be deposed by my lawyer. According to my lawyer, their expert on HUS praised my decision making and chart documentation and said that Jonathan was fortunate to have recovered from such a serious HUS episode. Following this deposition, I was dropped from the suit. Later the suit was settled before going to court. As often happens, the costs of a settlement were comparable to the costs of going to trial.

Thirteen years after Jonathan's hospitalization, I attended a function and saw Mark and Talia. We talked about the lawsuit. Mark said that he and Talia appreciated everything I had done to save Jonathan's life, but they needed to file the suit to ensure they would be able to afford additional hand surgeries. I knew Mark was right to be concerned about Jonathan's future health expenses and what might happen when he needed additional surgeries and therapy for his hand. Many types of insurance policies had exclusions for pre-existing conditions. If Mark became unemployed or changed jobs, he could lose insurance coverage for his family.

Later that same day, while walking in a park, I recognized a boy playing with a tennis ball. I glanced at the boy's left hand and saw he was missing part of his thumb and first two fingers. Jonathan seemed to recognize me as well, so I approached him to see how he was doing. He asked me to play catch with him.

"Aren't you Dr Berman who took care of me in the hospital?"

"Yes, I am . . . it's good to see you've done so well, Jonathan."

We started to throw a tennis ball back and forth. He used his unaffected right hand. "I feel fine, but I didn't like the surgeries on my hand. They hurt a lot."

"I'm sorry to hear they were so painful, but your hand looks pretty good."

"My hand doesn't look good, and kids tease me about it all the time."

In an angry accusatory tone he added, "Why did you do this to me?"

I felt a surge of hostility and wanted to turn and walk away. But I realized he had probably overheard many conversations about his hospitalization and my role. He was obviously upset about his hand. I wondered if I could help him understand what had happened.

"Jonathan, I can see you're angry at me, but I was trying to save your life. Sometimes when you try to save someone who is very sick and may die from their sickness, other things called complications happen. We don't know they will happen or want them to happen. They just happen."

"Are you saying God did this to me?"

I explained that I thought God wanted him to live and had helped him get better. I told him that I thought he was a very special boy and had wonderful parents.

"I'm glad I was able to help, but you and God did it. You were strong and wanted to live, and with God's help, you did. Perhaps your hand is a sign of your strength. Don't let the teasing bother you."

He then became a more typical 13-year-old who wanted to show me his baseball skills. After a few more minutes we parted. The encounter seemed to provide a sense of reconciliation for both of us, addressing his anger about his hand and mine about the lawsuit. In eighth grade, Jonathan called me to ask if I would help him do a science project on HUS.

I still ask myself how I could have acted differently to avoid the lawsuit. Should I have pulled Jonathan's brachial line as soon as he arrived even though he may not have been stabilized? Being sued hurts even when we haven't made bad decisions, provided standard care, or failed to communicate with patients and their families. In the absence of guaranteed health insurance, families such as Jonathan's will continue to find it necessary to sue their health providers, even when responsible medical decisions were made. For me, the experience of caring for Jonathan highlights 2 needs: the personal need to share his story with colleagues and enlist their support, and the professional responsibility to advocate for universal health care coverage.

This is excerpted from Dr Berman's book which will be published in 2008—*Getting it Right for Children: Stories of Pediatric Care and Advocacy*. Steve Berman. Elk Grove Village, IL: American Academy of Pediatrics; in press.