

Being a Father and a Doctor

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[N A J Med Sci. 2009;2(3): 100-101.]

When your 18 year old daughter becomes seriously ill, should you help the medical team or be uninvolved in her care? This is the dilemma I confronted in June 2008. My daughter was suffering from mysterious symptoms. It all started on a Tuesday morning the week of her 18th birthday. We were driving to my office together and she said something odd “Dad, my vision is blurry” Diana always had normal vision unlike her myopic Dad and brother. Fortunately, the ophthalmologist in our building was available that morning so she went for an exam that showed no significant abnormalities. Then she started to develop severe back pain. She appeared very restless and uncomfortable and started to cry. I suggested she go to the emergency room or see her pediatrician. She couldn’t drive so her boyfriend picked her up at the office and took her to the pediatrician. The pediatrician ordered some blood tests and sent her to another ophthalmologist that believed she had uveitis. Not realizing how sick she actually was as she left the office, I remember answering an inquisitive coworker about her problem that day saying she was a “drama queen”. This comment was not intended as a pejorative remark to her character, just recognition of her occasional penchant for being an emotive teenager at times when facing stressful situations. Those spoken words still haunt me today. Things got much worse. That night my children and I watched the Celtics – Lakers final in the family room. Diana continued to be very uncomfortable writhing in pain. Since her pain was not responding to high dose ibuprofen at all I allowed her to take some hydrocodone. She continued to show signs of discomfort as she dozed off and on during the game. Then on Wednesday and Thursday her symptoms were much milder. On Friday, the same problems happened again, severe back pain and blurry vision. It was her eighteenth birthday. Ironically, it was my practice manager’s wedding night too. We took her to our emergency room that afternoon. Surely, I thought, we would get her evaluated and comfortable and still make the wedding. How naïve I was. Poor Diana got her lumbar puncture at 5pm, the same time as the wedding ceremony. When the emergency physicians later explained

that she had meningitis, I was dumbfounded. She barely complained of any headache. I hoped she had viral meningitis as I knew this would portend the best prognosis. She might recover and be well again in several days. My optimism soon faded. The admitting medical team suspected possible Lyme meningitis as she had an indeterminate screening test. I did have one patient once with CNS Lyme disease, but really limited experience with this. I cross-referenced her symptoms: blurry vision and severe muscular pain, an unusual combination. Because of the uveitis, I did some independent reading and found rheumatologic conditions could also present with meningitis and uveitis. I decided to approach the medical resident and suggested a rheumatology consult just in case the Lyme disease diagnosis was incorrect. I’ll never forget the true kindness and appreciation of one of the medical residents. She was impressed that I had done so much reading to investigate all of the possible causes for the meningitis. I understood her completely: she certainly didn’t have a thriving, beautiful, talented 18 year old daughter who was now strangely ill with meningitis and writhing around in a darkened room. High doses of hydromorphone and lorazepam only allowed brief periods of rest interspersed with screaming pain and suffering. Welcome to Hades. During the second of her four admissions, the rheumatology fellow came to the room and told my daughter and wife that she had CNS lupus. My wife was angry and distraught. She (and I to a lesser extent) had kept vigil at the bedside for about 2 weeks seeing minimal improvement in her condition. The medical team continued treating her for CNS Lyme disease. Ultimately the rheumatology fellow would be vindicated. Diana turned out to have Primary Sjogren’s syndrome. The puzzle would unravel very slowly. It actually totaled over four weeks of hospital stays, several dozen physicians, and multiple specialists and took 9 months to solve. The first episode of illness took about 4 months to resolve.

My involvement as a doctor would both help and hurt her situation. The key clue to her diagnosis actually was present in her initial admission. She had self treated for cystitis with trimethoprim/sulfamethoxazole that Tuesday morning of her first week of illness. There were other clues that I pieced together for the rheumatologist. She had had an unusual parotid swelling and pain in January of 2008 that was diagnosed in the ER as a salivary stone (I thought it might have been viral). Unfortunately I did more than piece together history, I created it also. Diana was suffering a painful paronychia on her toe in November. The college health clinic advised warm soaks. She called feeling

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desperate for relief and I relented to prescribe some antibiotics. She now had a new primary care physician but often preferred to request help from me for convenience in urgent situations, especially now that she was calling from her campus in Western Massachusetts. She had a drug allergy to ceftriaxone, suffering fever, rash, and rigors early in the course of her illness so I did not want to use any type of beta lactam antibiotic. I suggested taking the trimethoprim/sulfamethoxazole again, especially since she had prolonged recent hospitalizations and might harbor MRSA. It turned out to be a fateful choice. When she filled the prescription several days later, she had the same problem again. Blurry vision, then severe muscle aches, headache and meningitis. At this point, we knew she must not have CNS Lyme. The rheumatologist cautioned she should not take sulfa or NSAIDs again due to the possibility of drug reaction. There was no firm diagnosis at this time, but a rheumatologic condition was still suspected. She had a high titer ANA positive test, but confirmatory tests for lupus were negative. She had high titer positive tests for SS-A and SS-B for Sjogren's syndrome. I reasoned this must have been the cause of her recurrent meningitis. I had carefully questioned her. "Diana did you take any NSAIDs or sulfa drugs prior to your first hospitalization in June or since or suffer any similar reactions". Her answer was essentially "no, I did not". Later on it became clear, she likely was very weary from all of the illness and had a temporary lapse in memory about the use of trimethoprim/sulfamethoxazole in June 2008. Much to my dismay, she would have another episode with another desperate call from campus. The toe surgery for the paronychia had failed and the toe infection returned. I thought perhaps I should use azithromycin. But Diana had confirmed no use of trimethoprim/sulfamethoxazole in June 2008. No MRSA had been found but she was clearly at risk with her multiple hospitalizations. So I prescribed the

trimethoprim/sulfamethoxazole again believing her history from June 2008 that no drug had been taken. Unfortunately, the same stereotyped reaction occurred: initially blurry vision, then severe myalgias and headache. She had another 1 week hospitalization and 4 months of recovery. In February, she had recurrence of parotid swelling and that seemed to clinch the diagnosis of Primary Sjogren's Syndrome. It appears the reason for her severe reactions is likely due to this underlying autoimmune disorder. She had a "serum-sickness" like reaction also known as type III hypersensitivity reaction due to immune complexes. I have to believe it is very rare because I have never seen it before nor heard of other recent cases. I imagine several decades ago it was commonplace before our improved type and cross matching techniques for blood product administration.

My final impressions about this horrible odyssey are as follows : 1) being a father and watching severe suffering in your child would be even worse if you could not offer some of your expertise to assist your caretakers. I am very grateful for the opportunity to provide suggestions to her medical team. 2) beware of the seduction of family and friends to provide medical care outside of the normal boundaries of your work and patient-doctor relations. I actually have successfully deflected more than 90% of all requests from family members over the years. My typical response is to listen, discuss, and suggest discussing with your own doctor. Even in my daughter's case, I made many efforts to redirect her to her primary care doctor or specialist at key junctures in her treatment.

You may wonder why I would bother with this diatribe. Well, I think we all need some catharsis now and then.