MELAS Syndrome Patient History

Volume 2.0

This publication contains an abbreviated version of my wife's medical history as an individual with the full form of the MELAS Syndrome, a mitochondrial disorder.

(Some of the symptoms and clinical signs of the MELAS Syndrome discussed in this publication may be applicable to other mitochondrial disorders.)

Karen Ann Jackson 1956-1992

This material was created, (and/or) assembled and edited by Michael J. Jackson
And is available in PDF format at the following URL:
Jackson Family's Website: http://www.melas.org

My thanks go to the doctors, nurses, researchers, medical personnel and facilities, support groups and organizations, "mito" families, and individuals that all contributed some part in making it possible for me to produce this publication. My thanks go also to my parents for their support, comfort, and encouragement during my wife's illness and when she finally succumbed to the MELAS syndrome.

I also have a special thank you for my son, Michael.
Thank you for loving Dad so much, helping me to adjust to Mommy going Home, and for helping me with this project and the MELAS Online Network (closed August, 1998.)

My most deeply felt thanks goes to my wife Karen Ann for so bravely enduring her illness, and for her encouragement to make her suffering into something that would help other families and individuals with mitochondrial disorders.

… and, without God's grace and mercy this publication would never have existed.

- Michael J. Jackson

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This is Mike, Karen's husband. A good deal of this history is based on a medical history of Karen done at Emory University's Neuropsych Unit in Atlanta, GA. I was interviewed, and my account recorded for use in Karen's treatment. I have never met such a dedicated, caring and professional team of psychiatrists and psychologists in the time before or during the ten-year period of Karen's illness, or to the present day as the marvelous team at Emory's Neuropsych Unit. Thank you, Folks! (And Mike still has the bear with the Indian name <smile>). This publication will carry the history to the present.

Karen was born in 1956 by NVD with out complications. Her childhood medical and neuropsychiatric histories are unremarkable except for a possible undocumented hearing deficit. According to Karen, her mother complained often about having to always call for her twice before she would turn. Karen experienced brief and infrequent blackout spells while she was in college, although it is unclear whether or not they were seizures. We met at college in 1975.

I didn't recognize any medical or neuropsychological problems until three years after we were married in 1977, which was in 1980 when Karen began to complain of fatigue, but I do recall Karen as being always poorly coordinated on her feet.

During the 1980-81 school year Karen would come home from her day of teaching school increasingly exhausted, which was unusual since she had always been energetic. Also that year she developed momentary episodes of visual problems. Bilaterally her visual fields would fill with black spots. At first this was just bothersome, but in time the episodes progressed to "brown outs" with some mild accompanying dizziness and vertigo. Within a year after the initial episodes of visual problems, the episodes progressed to grand mal seizures. They appeared as sudden onset "black spots" that increase in size and number until Karen's entire visual field would go black. Then she would grow dizzy and have a seizure. The aura of black spots would continue to precede her seizures for the years to come.

From 1981 to 1984 Karen exhibited no obvious intellectual or memory problems. She was "normal" except for a mild to moderate hearing deficit which may have been progressively getting worse; weakness and increasing ataxia; and a seizure disorder that was difficult to control. I didn't notice any behavioral or personality changes, although I'll admit that she was perhaps a bit more irritable during this period.

Control of Karen's seizures was originally tried with Dilantin. Over time she developed increasing weakness, ataxia and severe confusion accompanied with hallucinations. Initially there was some question as to whether this was the progression of her disorder or a side effect of her Dilantin, since there did seem to be a relationship between the Dilantin and these symptoms. Karen also began to develop bizarre delusions that progressed to hallucinosis. These experiences included depersonalization, out of body experiences and paranoia.

For example, while in the hospital Karen became horrified by her I.V. because she was convinced that every patient was connected to the same I.V. and that tubing "was everywhere." During another severe delusional episode Karen could not drink from a soda bottle because she thought it was part of her. These symptoms ceased with discontinuation of Dilantin. Tegretol was used next, but similar problems with weakness, atoxia, confusion and later hallucinosis developed. In addition, she became leukopenic. Phenobarbital was next used with good control even though at times fairly heavy, sedating doses were necessary to prevent seizures.

In 1978 Karen developed hepatitis (caught from a Sunday school student who contaminated the ladies room. Six church ladies got it).

From 1984 to 1986 no seizures were witnessed. Still didn't notice any behavioral or personality changes. As to the question of a progressive dementia, I don't recall any significant change in cognitive function or memory, although her energy level and strength did drop off slightly during this period. Forgetfulness regarding the location of household objects may have increased.

The year 1987 was marked by a dramatic increase in Karen's seizure activity. She had severe recurrences of status epileptius, being left comatose after the third time for three to four weeks. (From this point on, with my son at around ten months old, I was functionally a single parent. This really complicated caring for Karen in her rapidly degrading condition.)

Karen never fully recovered from this coma. Prior to the onset of coma, there was a step-wise decline in cognitive function and personality with each episode of status epileptius. Waking up shortly after the first two episodes, her ability to follow a normal conversation, to understand formal language and multi-part tasks had decreased. Karen's memory also suffered so that she needed more constant reminding. With regard to personality, Karen began to intermittently lose her ability to focus on others. She was often unable to understand what I was feeling, and she often had difficulty sympathizing and empathizing with others while she became increasingly more "lost" within her own world. She became unable to perform childcare responsibilities associated with our infant son.

Following the third episode of status with coma, Karen was very slow to recover even basic functions. It took eight months for her to be able to rejoin us at home. Her ability to walk varied and she often remained confined to her wheelchair. Significant global muscle atrophy left her increasingly weak and debilitated. During this recovery process Karen may have been slightly depressed. While she retained much of her sense of humor during this period, her affect was occasionally inappropriate. Unfortunately, the same was not true of her cognitive functions. Again, a large step-wise reduction in abilities occurred during this period.

Karen was now totally dependent upon the care of others, and myself, and she lost any sense of what was going on in the world around her. When asked a direct question, she would answer "indefinitely," speaking very loosely until broken-off or left alone.

Following the coma Karen began to experience marked mood swings. These behavioral changes began while she was hospitalized at the Medical College of Georgia and continued throughout her nursing home stay and after returning home. Up until this time her only personality/behavioral changes had been social withdrawal and loss of understanding the outside world. The outgoing, extroverted, giving personality had become selfish, introverted, and unable to comprehend the needs of others. Her mood swings included a whole new area of difficulty.

Karen's mood could vary from day to day as could "Dr. Jekyll and Mr. Hyde." Some days she would act like an angry, spoiled, and selfish child. At first this would not last more than a day and would be characterized as being demanding and verbally abusive. Then the change became more frequent and lasting days, often ending in hospitalization secondary to violent uncontrollable behavior. A typical severe mood swing would begin with agitation and defiance. Karen would be verbally abusive and threatening to me.

For example, she would threaten to have the state take our son away from me. She would also adamantly refuse to eat or drink. Nausea and vomiting could also occur with a higher level of agitation. The episodes of vomiting increased in frequency and severity; and were usually accompanied by severe headaches. Within one or two days she would become increasingly irritable and confused, perhaps related to the dehydration. She would become completely unreasonable, often screaming incoherently.

Finally her condition would deteriorate to hysteria and violence against anyone or anything, thus requiring restraint and hospitalization. After hospitalization, usually within a 24 hour period, she could be "herself" again.

These mood swings made her management at home increasingly difficult and stressful. Karen was hospitalized in the Neuropsychiatric Unit at the Wesley Woods Hospital in Atlanta, Georgia from 4/25-5/3/89. Observations during Karen's hospitalization revealed that the agitation and defiance stage lasts for longer and longer periods of time, occasionally for more than ten days, with only brief (<20 hours) respites every five days or so.

Karen's condition was complicated by "stroke-like episodes." In February 1988, Karen suffered a sudden drop in visual activity bilaterally throughout her visual fields. Karen went rapidly downhill after this admission. Karen also had several episodes of unilateral weakness; numbness and tingling lasting only hours with seemingly full recovery. These later episodes were not clinically documented, a disappointment since this would probably have added to the knowledge gained from Karen's bout with the MELAS Syndrome.

Our son was born in 1987, the same year that Karen was finally diagnosed with the MELAS Syndrome. From the time that Mike was ten months old and on, Karen was unable to care for him. Karen spent most of 1988 in a coma and recovering from it. 1989 was spent at home with us, with several visits a week from a local home nursing program.

I had to quit working for six months in order to get our resources low enough to qualify Karen for SSI (disability, she didn't qualify for SS). Karen was running medical bills in the neighborhood of $100,000.00 to $200,000.00 per year by this time. We used Welfare and the SSI for expenses. Medicaid and the State (GA) picked up the tab for Karen's medical bills. Kept trying to build a business in order to stay self-employed. Thought that it would be better for being a single parent later. Turned out to be hard to do without any money to invest in it. (The third quarter of 1997 is the first in years when I've been able to pay our monthly bills on time. Whew, long time. It'll be touch and go for a long time yet, though.)

In 1990 Karen's violent spells became more frequent. I would wake up in the night to find her beating the living daylights out of me. Had scars for a while. I began to be afraid that Karen would unintentionally hurt our son, or worse, that I would loose it and swing back. Finally gave into the doctors, and had Karen placed in a nursing facility (nice name for a nursing home). During 1990, Karen would mostly recognize us, but not always. Sometimes she thought we were dead, sometimes that I had died and our son had grown, gone to college and married. That year she wasn't much more than 10 or eleven mentally. We would bring her home on weekends, or stay there with her all day. I would eat lunch with her during the week when I could.

1991 was worse. In those last three years, Karen let me hold her twice. Nice moments, but the rest of that time, when she did recognize me, she would become extremely agitated and rant and threaten me for placing her in the home. She was mostly out of it. She continued to deteriorate and lose body mass.

1992 was the last year that Karen was with us. Very little response to stimuli and she didn't recognize anyone. On December 4th, Karen developed a high fever and was gone in less than an hour. Eleven days later, we would have had our 15th wedding anniversary.

Most of what is going on with us now is available at the Jackson Family's Website on our personal pages.

- Mike Jackson
March, 28, 2000