



Great Expectations

Pregnancy and Childbirth with Neuromuscular Disease

by Margaret Wahl, Amy Labbe and Miriam Davidson

Having a baby can be the most thrilling and rewarding experience of a woman's life, yet it also can be fraught with fear and uncertainty. This is especially true for women with neuromuscular disease.

This special MDA report takes a look at the issues that arise for expectant mothers with [muscle disease](#) and finds that, with proper care and planning, these women are usually — although not always — able to have successful pregnancies and give birth to healthy children.

The report contains information from the July-September 2010 issue of MDA's Quest magazine, as well as additional information not found in the print magazine.

Contents include:

Caution, Preparation and Teamwork Lead to the Best Pregnancy Outcomes in Women with Neuromuscular Diseases	2
Disease-Specific Complications (chart)	7
Good Advice from Women Who've Been There	11
Medication Complications for Pregnant Women with Neuromuscular Disease (chart)	17
Personal Stories	
Diagnosis Later in Life	3
Baby Born with Challenges	4
Scared and Worried	6
Achieving All Her Goals	9
Worth the Effort	10
The Nurse Called It 'Fred's Ataxia'	12
A Turn of Fate	15
Surprise Pregnancies	16

In addition, listen to a [podcast of an interview](#) with neurologist Emma Ciafaloni and obstetrician Eva Pressman by going to quest.mda.org/podcasts.

Caution, Preparation and Teamwork Lead to the Best Pregnancy Outcomes in Women with Neuromuscular Diseases

When Emma Ciafaloni was preparing to become a neuromuscular disease specialist in the 1990s, and even when she directed the MDA neuromuscular disease clinic at Duke University in the early 2000s, she was struck by the lack of information she could offer patients who wanted to become pregnant.

“I really have an interest in women and [neuromuscular diseases](#), in what we can do better for them in dealing with their neuromuscular disease and also with pregnancy,” says Ciafaloni, now at the University of Rochester Medical Center, where she sees patients in the [MDA clinic](#) and has received [MDA research](#) support.

“I’m very interested in how to best care for patients. I’m not the one who’s going to discover the treatment for [FSHD \[facioscapulohumeral muscular dystrophy\]](#) or [myotonic dystrophy](#), but I’m very interested in good standards of care.”

Ciafaloni’s interest ultimately led her to collaborate with several colleagues in the departments of neurology and obstetrics and gynecology to conduct a study of pregnancy in women with FSHD. The results were published in 2006 in the journal *Neurology*.

The researchers administered questionnaires to, and reviewed the medical records of, 38 women with FSHD. On the whole, pregnancy outcomes were good in this group, although the rates of operative deliveries (Caesareans and forceps deliveries) and babies with low birth weights were higher than the national average.

About one in four of the women sur-

veyed reported worsening of FSHD symptoms that for the most part did not resolve after child-birth. The most common problems were, in order of frequency: worsening of generalized weakness, frequent falling, difficulty carrying the newborn due to worsening shoulder weakness, worsening or new-onset pain, and difficulty carrying the newborn due to worsening of leg weakness.

Despite some child-birth complications and possible permanent worsening of their FSHD, 90 percent of the women said they would choose pregnancy again.

Eva Pressman, director of maternal-fetal medicine at the University of Rochester, collaborated with Ciafaloni and others on the FSHD study. “The important thing about our study was that the outcomes were really good,” she says. “It’s important for patients to know that just because they have an underlying disorder, that doesn’t mean that pregnancy is contraindicated. With appropriate monitoring and going into the pregnancy with your eyes open as to what the risks might be, I think most patients can be quite successful.”

But not all neuromuscular disorders, let alone all women or all babies, are the same. And even with conditions like FSHD, where the odds of having a successful outcome appear to be good, there are steps women can take to improve them.



An echocardiogram to evaluate heart function prior to becoming pregnant can help with decision making.

First, some red flags:

Cardiac involvement

Cardiac involvement can occur in many neuromuscular diseases and, if it’s severe, can be a major problem for women considering pregnancy.

Pregnancy leads to a significant increase in cardiac output, as well as a 50-percent increase in blood volume, so the heart does “much, much more work,” says Pressman. “And if you go into the pregnancy with a heart that’s functioning less than optimally, it will clearly deteriorate over the course of the pregnancy. That can lead to heart failure in the mother and endanger both the mother and the fetus, depending on when in the pregnancy those issues occur. The most common time for heart problems is the end of the second trimester or the beginning of the third trimester.”

Pressman strongly recommends that patients who are at risk for cardiac

dysfunction have echocardiograms before pregnancy to evaluate their heart function and “really consider not becoming pregnant if they have significant cardiac dysfunction.”

And if they're already pregnant and want to continue the pregnancy despite their heart problem? “Then you do the best you can,” Pressman says. “You can manage some of it with medications, you can keep their fluid status at an optimal level, and we can usually get them through to the point where the baby is viable. But often we deliver these patients early, to minimize the stress on the heart.”

Weak respiratory muscles

Weakness of the respiratory muscles can also be a problem for pregnant women with muscle disease. As the pregnancy progresses, the work of breathing becomes harder, so any

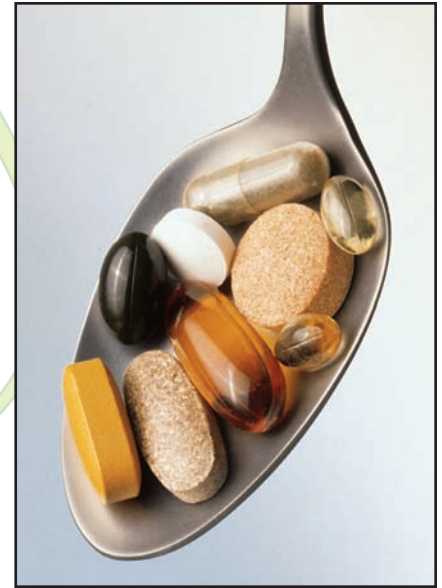
existing impairment can become more problematic.

“We see women who have reasonable respiratory function prior to pregnancy that deteriorate as the pregnancy progresses,” Pressman says. They generally recover to their pre-pregnancy level after delivery, she notes, but they may require extra support (such as noninvasive assisted ventilation) during the pregnancy.

Says Pressman, “We often end up delivering a little bit early, because the respiratory impairment only gets worse towards the end of pregnancy, and sometimes the safest thing is to not be pregnant anymore.”

Unstable autoimmune disease

It's critical that women with autoimmune disease are medically stable and on a stable regimen of medications for at least six to 12 months before



Immunosuppressant medications that aren't corticosteroids can pose some problems. See “Medication Complications for Pregnant Women with Neuromuscular Disease” on page 17.

they try to get pregnant, says Hannah Briemberg, a neurologist and assistant professor in the neuromuscular dis-

Diagnosis Late in Life

**Kathy Rivera, 58
Tucson, Ariz.
type 2 Charcot-Marie-Tooth disease**

Like many people with Charcot-Marie-Tooth (CMT), Kathy Rivera didn't

realize she had a neuromuscular disease until relatively late in life. Called “clumsy” as a child, she wore special shoes to keep her feet facing forward, but didn't receive an official diagnosis of CMT until age 36.

Rivera received almost no medical care growing up, which partially explains the late diagnosis. She moved a lot, spent time in foster care and was raised primarily by an uncle. Her symptoms were shrugged off as “growing pains.”

Rivera married and had her first child, a healthy girl, at age 27. For reasons probably unrelated to CMT, Rivera had high blood sugar during the pregnancy and, during the birth, her cervix (entrance to the uterus) didn't open. “I tried to give birth



Kathy Rivera, her husband, Tom, and their two children, in 1985

the natural way and almost lost her,” Rivera recalled. “I had to have an emergency C-section [Caesarean], and she had to be resuscitated.”

During her second pregnancy at age 33, Rivera's still-undiagnosed CMT noticeably worsened. Her legs were so sore when she woke up that she couldn't get out of bed. “We put the bed next to the wall so I could climb up the wall to get to a standing position. I thought, ‘I'm not having any more kids.’”

As with her first pregnancy, Rivera had high blood sugar. Her son was born healthy via a planned C-section.

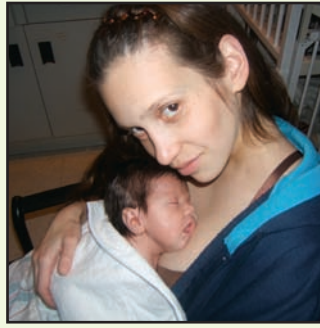
Rivera's symptoms continued to worsen, and it was her daughter who convinced her to see a neurologist. “My 6-year-old said to me, ‘Mom, why don't you pick up your feet?’”

Today, Rivera wears braces on both legs and no longer works. She loved her job as a bakery manager at Safeway, but her doctor advised her to quit. In recent years, Rivera has developed nerve compression requiring surgery in both wrists (likely related to CMT) and spinal cord inflammation (which may or may not be related to CMT).

Rivera believes her now-adult daughter may have CMT too, although she's never been tested. Her daughter decided not to have children, but instead adopted a little girl. In an ironic twist, Rivera's adopted grandchild has muscle problems and is being tested at the MDA clinic in Tucson for possible neuromuscular disease — receiving the care and attention that Rivera never did.

Baby Born with Challenges

Liz Trumpy, 39
Levittown, N.Y.
type 1 myotonic dystrophy



Liz Trumpy and her daughter, Kelly, who was born in January 2009

Liz Trumpy was an active, apparently healthy New York City police captain who worked out with weights, ran a marathon and participated in mini-triathlons. Married to another police officer, she went to the gym regularly throughout her pregnancy, even on the day before she gave birth.

“If Kelly had been born without health problems, I would have said I had a picture-perfect pregnancy,” says Trumpy.

But, to Trumpy’s shock and that of her doctors, her baby had **congenital myotonic muscular dystrophy**, the most severe form of the disease. Congenital MMD can cause problems with the heart, lungs, and cognitive functioning, in addition to muscle weakness and wasting.

Kelly had a rough start. She was two weeks late, so doctors induced her birth. When labor failed to progress, and Kelly’s heart rate began to drop, a Caesarean was performed. Kelly didn’t make any sound at first, and then, after a few moments, managed only a weak cry.

Kelly had very low muscle tone, fluid in her lungs and a club-foot (a foot that’s abnormally turned inward, which can occur

from muscle weakness). She almost didn’t survive her first night and had to be rushed to a hospital that had a neonatal intensive care unit, where she spent six weeks before her parents could take her home.

It was only after Kelly’s birth that Trumpy learned she has the **adult-onset form of type 1 myotonic dystrophy (MMD1, also sometimes called DM1)**, a widely variable form of muscular dystrophy that can cause congenital MMD in infants of affected women. (Men with MMD1 can father children with congenital MMD1, although it’s much more likely to occur when the mother is affected. Each child from a mother with MMD1 has a 50-50 chance of having MMD1, although not necessarily the severe, congenital form of the disease.)

Trumpy retired from the police force to take care of Kelly, and their days are full of visits with therapists, including speech, physical, occupational and educational. At 15 months, Kelly can walk using a walker, and talks baby talk, although she has yet to say her first word. She’s learning sign language. Her heart and lungs are all right.

Trumpy wishes she’d known about her MMD so she could have been better prepared for Kelly’s birth. She would have chosen a different hospital and a doctor more familiar with the kinds of issues that can arise for pregnant women with neuromuscular disease. As it was, she felt some members of her medical team were cold to her, as if they blamed her for Kelly’s condition.

Trumpy says she prefers to look forward, not back, and now that she has the support she needs, she’s able to focus on providing Kelly the best future possible. “She is our gift,” Trumpy says.

eases unit at the University of British Columbia (Canada) in Vancouver.

The neuromuscular autoimmune diseases in MDA’s program — **poly-myositis, dermatomyositis, myasthenia gravis** and **Lambert-Eaton myasthenic syndrome** — are generally treated with immunosuppressive drugs and sometimes other medications.

In a paper she published in 2007 in the journal *Seminars in Neurology*, Briemberg found that pregnancy does not appear to alter the long-term outcome of myasthenia gravis (MG) but that, if the disease is not yet stable before conception, there is more risk that it will worsen during pregnancy.

A phenomenon that can occur in MG but does not seem to occur in other

autoimmune diseases is the temporary transmission of the disease to the baby, causing him or her to be born floppy and possibly with swallowing or breathing difficulties. (The phenomenon occurs because the antibodies the mother’s immune system produces that weaken her own muscles can cross the placenta and weaken the baby’s muscles, at least temporarily.)

That doesn’t seem to occur as often as it used to, Briemberg says, something she attributes to better MG care and mothers whose disease is better controlled during pregnancy.

Women with polymyositis or dermatomyositis also need to have their disease under control, preferably before getting pregnant. Briemberg found

that women with myositis who were in remission at the time of their pregnancy and delivery did not appear to be at risk for obstetric complications. However, she found, women with active disease at the time of their pregnancies had an increased incidence of spontaneous abortion, premature delivery and low-birth-weight infants. The highest risk was associated with new-onset disease during the first trimester of pregnancy. In these women, the rate of fetal deaths was very high (although the number of women studied was small).

Although there are risks associated with the use of disease-controlling medications during pregnancy, Briemberg advises that these risks are, in general, preferable to the risk of

being pregnant and having an uncontrolled autoimmune disease.

Corticosteroid drugs, such as prednisone, are often prescribed for autoimmune diseases, and generally appear to be fairly safe during pregnancy.

“Prednisone doesn’t cross the placenta very well,” says Eva Pressman, “so it’s one of the safer medications to use during pregnancy, because most of it is going to the mother, with very little of it getting to the baby.” It’s not entirely without risk, however. Prednisone and related medications can interfere with the growth of the fetus and have been associated with premature rupture of the amniotic sac, possibly by interfering with collagen formation, she notes.

Pressman adds that it can be impossible to determine whether problems are from the effects of prednisone or the effects of the mother’s underlying disease.

Another issue with corticosteroids is that, if they’re given for a long period of time, they can suppress the function of the adrenal glands, which normally pump out high levels of the stress-coping hormone cortisol during labor and delivery. Therefore, in general, doctors recommend that women who have been on prednisone or other corticosteroids be given intravenous corticosteroids (hydrocortisone) during labor and delivery.

Immunosuppressant medications that aren’t corticosteroids pose different types of concerns. See the chart [“Medication Complications for Pregnant Women with Neuromuscular Disease”](#) on [page 17](#).

None of these medications have been systematically studied in pregnant women, Briemberg notes, so most of the data comes from animal experiments or data collected from women who happened to get pregnant while taking one of them.

“At this point, the clinical data and

experience suggest that prednisone, azathioprine and IVIG [intravenous immunoglobulins] are unlikely to pose any significantly increased risk of fetal malformation,” Briemberg says. “There is not enough data on other immunosuppressive medications to know if they are safe in pregnancy, so most clinicians will recommend coming off these other medications prior to trying to conceive.”

Myotonic dystrophy — a special case

Most neuromuscular diseases affect mainly the voluntary muscles (in the limbs, trunk, head, face, and swallowing and breathing structures). The heart, though not a voluntary muscle, also is affected in many neuromuscular diseases.

But [myotonic dystrophy \(MMD\)](#) affects not only the voluntary muscles and the heart, but the involuntary, or “smooth,” muscles that line the hollow organs, such as the gastrointestinal tract, urinary tract, uterus and vagina.

Abnormalities of uterine and vaginal muscle function (either weakness or myotonia, the inability to relax muscles) can have severe adverse effects on labor and delivery.

“If the uterus is actually affected, then labor may not progress well,” Pressman says, “and you may not be able to have a vaginal delivery. We can try to alter that with oxytocin [a labor-stimulating hormone] or other medications, but if you can’t make the uterus contract, then you would need a C-section [Caesarean] to deliver.”

In addition, people with MMD are especially sensitive to pain-relieving medications and can have abnormal reactions to anesthesia, so there are additional worries on this account.

A very severe form of MMD called congenital MMD can occur in offspring of women with type 1 MMD who may themselves be minimally affected. So



Kelly Trumpy, born with congenital myotonic dystrophy, is learning to walk using a posterior walker.

far, this phenomenon has not been seen in type 2 MMD. (Type 1 MMD is caused by an expansion of DNA on chromosome 19, while type 2 MMD, a similar disease, is caused by an expansion of DNA on chromosome 3.)

Babies with congenital MMD can be born very floppy, with respiratory impairment and sucking and swallowing difficulties, for which the obstetric and pediatric teams must be prepared.

Normally, babies swallow some of the mother’s amniotic fluid, the watery substance that surrounds the baby in the uterus. But a baby with congenital MMD can have so much swallowing impairment that excess amniotic fluid accumulates, further endangering mother and baby.

Excess amniotic fluid, called “polyhydramnios,” can cause premature rupture of the membranes and premature onset of labor, sometimes before the baby is ready to be born. If the membranes don’t rupture prematurely, the uterus can become so distended that the mother’s breathing is impaired and blood vessels can be compressed. Excess bleeding after delivery (postpartum) is also associated with an overdistended uterus during the pregnancy.

“If you have a baby with limited

swallowing ability and you have polyhydramnios, and you've overstretched the uterus because of that, then even a normal uterus doesn't contract well," Pressman says. With a uterus affected by MMD, she notes, "your risk of postpartum hemorrhage is much higher."

Planning Ahead: Five P's

Traditionally, obstetricians and midwives have thought about pregnancy, labor and delivery in terms of three p's: the *passenger* (baby), the *passageway* (the mother's bone structure and soft tissues of the birth canal), and the *powers* (the involuntary contractions of the uterus and the voluntary pushing efforts of the mother's abdominal muscles).

To this classical way of looking at things, two more p's can be added: *pain management* and *pregnancy-disease interaction*.

Almost any pregnant woman with a [neuromuscular disease](#) can have difficulty with at least two of the three p's, the *passageway* and the *powers*. And, if those difficulties are serious, they can affect the *passenger* as well.

In addition, pain management can pose some special challenges for some women with neuromuscular disease, and sometimes pregnancy can have some long-term adverse effects on disease progression.

Thinking of pregnancy and delivery in terms of the five P's can help women with neuromuscular disease plan ahead.

The passageway for the passenger

Several factors can affect the passageway.

"The size of the mother's pelvis is clearly important and can affect a woman's ability to have a safe vaginal delivery," says Eva Pressman.

David Colombo, a maternal-fetal med-

icine specialist at Ohio State University Medical Center in Columbus, has seen small "juvenile-type" pelvises in several of his patients with [spinal muscular atrophy \(SMA\)](#). He prefers to do a C-section in these cases.

Says Pressman, "It's not required to have a C-section just because of a small pelvic diameter, but it would be the kind of thing where you would have the discussion ahead of time, watch the progress of labor, and perhaps not wait through three or four days of labor before deciding enough is enough."

Sometimes, contractures (joints frozen in one position) in the hips, knees or spine can pose an impediment to delivery and may lead a physician to recommend a C-section.

Spinal curvatures — and sometimes the surgical procedures performed to

... continued on [page 9](#)

Scared and Worried

Aimee Chamernik, 40
Grayslake, Ill.
amyotrophic lateral
sclerosis (ALS)



Aimee Chamernik was 33 and just beginning her third pregnancy when she noticed she was having difficulty enunciating as she read to her son at bedtime. "I thought it was odd, but I chalked it up to being tired after caring for two small children all day with a third on the way," Chamernik says.

Chamernik mentioned the problem to her obstetrician at her next prenatal appointment, and was referred to a neurologist, who tentatively diagnosed her with [myasthenia gravis](#), an autoimmune neuromuscular disease, and put her on Mestinon (pyridostigmine bromide). But when her speech continued to slur and deteriorate, and she noticed some weakness in her right wrist, she sought a second opinion. Further tests ruled out myasthenia gravis, and Chamernik stopped taking Mestinon.

Throughout the rest of her pregnancy, Chamernik's speech continued to slowly worsen, she began to choke

more frequently, and her wrist weakness grew more pronounced. Chamernik suspected she had [ALS](#); unfortunately, she was right.

Although her ALS was not formally diagnosed until a year after her third child was born, Chamernik spent most of her last pregnancy feeling scared and worried.

"It's easier now to look back and realize that ALS had little, if any, impact on my pregnancy," Chamernik says. "However, at the time, I was awake late, late into the night a lot, worrying about my baby, crying about my other children possibly having to grow up without a mom based on what was happening to me, tormenting myself while researching the possibilities (which was a terrible idea, but one I couldn't seem to resist). I wish I hadn't wasted so much time worrying and crying — it didn't accomplish anything except making me lose sleep, and it didn't prevent my eventual ALS diagnosis."

Chamernik had no problems with the labor and delivery of her third child. "One thing I wish I had understood better and believed more strongly is that the changes that were happening to me had little or no impact on my baby," she says. "He grew and developed and hit every milestone, despite the weakening of my throat and mouth muscles. At the time, though, no amount of reassurance from doctors could ever truly allay my fears."

Disease-Specific Complications

Disease	Complicating Factors	Associated Risks
ALS (amyotrophic lateral sclerosis, or Lou Gehrig's disease)	generalized weakness and muscle atrophy; paralysis; respiratory insufficiency	<ul style="list-style-type: none"> risk of respiratory failure weakened muscles may decrease ability to push during delivery increased rate of delivery interventions*
carnitine deficiency (CD)	weakness in the hips, shoulders, and upper arms and legs; heart muscle weakness	<ul style="list-style-type: none"> decreased levels of carnitine caused by pregnancy can lead to irreversible muscle damage; deterioration can continue even after childbirth
carnitine palmitoyl-transferase deficiency (CPT)	episodic muscle pain, weakness and tenderness; in some women, smooth uterine muscle may be impaired	<ul style="list-style-type: none"> outcomes vary widely, from uncomplicated vaginal delivery to postpartum hemorrhage and emergency hysterectomy
central core disease (CCD)	episodic muscle pain, weakness and tenderness; in some women, smooth uterine muscle may be impaired	<ul style="list-style-type: none"> high risk of malignant hyperthermia if general anesthesia is used
Charcot-Marie-Tooth disease (CMT)	muscle weakness and atrophy; contractures; possible curvature of the spine	<ul style="list-style-type: none"> exacerbation of weakness (sometimes temporary, sometimes not) higher rate of abnormal fetal positioning significant increase in delivery interventions* increased risk of postpartum bleeding or hemorrhage
dermatomyositis (DM) and polymyositis (PM)	weakness and pain in the muscles of the hips, thighs, back, shoulders and neck; possible heart and respiratory problems	<ul style="list-style-type: none"> outcome of pregnancy is closely linked with mother's health. When the disease is active, there's a higher risk of spontaneous abortion, intra-uterine growth retardation, fetal death, premature delivery and low-birth-weight infants. When the disease is in remission, there appears to be minimal risk to mother and fetus. extreme muscle weakness may necessitate assisted labor and delivery* PM and DM may cause elevated blood levels of creatine kinase (an enzyme that leaks from damaged muscles) in the newborn for a few months after birth
facioscapulohumeral muscular dystrophy (FSHD)	muscle weakness in legs, abdominal and hip muscles	<ul style="list-style-type: none"> increased muscle weakness during pregnancy that may not resolve after childbirth; worsening or new onset pain reduced ability to push during labor, leading to increased incidence of delivery interventions* increased incidence of low birth weights
Friedreich's ataxia (FA)	loss of balance and coordination; weakness in the legs, arms and hands; muscle spasticity; skeletal abnormalities; cardiac abnormalities; possible diabetes or glucose intolerance	<ul style="list-style-type: none"> may aggravate diabetic or borderline diabetic condition cardiac problems may worsen; the heart may not be able to pump enough blood to the body and to the placenta, depriving both mother and baby of oxygen; heart deterioration may lead to heart failure
limb-girdle muscular dystrophy (LGMD)	weakness and atrophy of the muscles of the shoulders and hips	<ul style="list-style-type: none"> possible worsening of weakness, with some recovery after delivery higher risk of Caesarean section
multi-minicore disease (MMD)	muscle weakness in the trunk; respiratory problems; skeletal deformities; spinal curvature	<ul style="list-style-type: none"> increased risk of developing anesthesia-induced malignant hypothermia

Disease-Specific Complications

Disease	Complication Factors	Associated Risks
<p>myasthenia gravis (MG)</p>	<p>fatigue and some generalized weakness of voluntary muscles</p>	<ul style="list-style-type: none"> • clinical worsening during pregnancy and in the month following child-birth; subsequent pregnancies in the same woman may have different relapse patterns • risk factors that could cause maternal death: respiratory failure, cholinergic crisis (overstimulation of the neuromuscular junction due to an excess of acetylcholine), pre-eclampsia, and postpartum hemorrhage • weakness in muscles associated with pushing may necessitate assisted delivery* • postpartum exacerbations that often are sudden and associated with respiratory failure • high mortality rate in fetuses and newborns up to 4 weeks old • fetal complications including multiple joint contractures due to lack of movement during gestation, and increased risk of infant death due to lung underdevelopment • temporary transmission of MG from mother to baby, resulting in poor sucking and swallowing, generalized poor muscle tone and respiratory distress for several months
<p>myotonic dystrophy type 1 (MMD1, or DM1)</p>	<p>generalized muscle weakness and atrophy in voluntary muscles; smooth muscle abnormalities affecting the uterus and vagina; heart and respiration problems</p>	<ul style="list-style-type: none"> • higher rates of infertility in both women and men may preclude pregnancy • potential worsening of disease symptoms including muscle weakness and pain; these may disappear after delivery • increased risk of ectopic pregnancies; early spontaneous abortion; excess amniotic fluid; urinary tract infections in the mother; pre-eclampsia; placenta previa; premature labor/delivery; death of the fetus or newborn; postpartum hemorrhage • problems with the first stage of labor resulting from dysfunction of the smooth muscles of the uterus and vagina • deterioration of heart function, possibly leading to heart failure • worsening of respiratory problems • risk of adverse reactions to general anesthesia and pain-killing medications given intravenously during labor • about 25 percent risk of passing along a severe form of the disease, congenital MMD, to the child, if mother has MMD1
<p>myotonic dystrophy type 2 (MMD2, or DM2)</p>	<p>generalized muscle weakness and atrophy; heart and respiration problems</p>	<ul style="list-style-type: none"> • possible hastening of the onset of disease • worsening of disease symptoms including muscle weakness and pain; these may disappear after childbirth • deterioration of heart function, which may lead to heart failure • worsening of respiratory problems
<p>paramyotonia congenita (PC)</p>	<p>episodes of prolonged muscle contraction and/or weakness</p>	<ul style="list-style-type: none"> • possible worsening of PC symptoms
<p>spinal muscular atrophy (SMA)</p>	<p>muscle weakness in the shoulders, hips, thighs and upper back; respiratory muscle weakness; skeletal deformity; contractures; curvature of the spine</p>	<ul style="list-style-type: none"> • worsening of weakness and increased fatigue • increased risk of urinary tract infections • spinal deformities or spinal fusion may interfere with the use of spinal or epidural anesthesia • mother's contractures may complicate delivery and/or anesthesia • increased incidence of premature delivery • possible loss of sensation or paralysis in the mother due to the pressure of the growing uterus on an abnormally shaped or fused spine

*“Delivery interventions” refers to assists used during the birth process such Caesarean delivery (C-section), forceps and vacuum assist.

Achieving All Her Goals

Stacy Wiparina, 35
Dayton, Ohio
type 2 spinal muscular
atrophy

Stacy Wiparina has never walked — never even crawled — but she's achieved all the goals she set for herself as a child. "One was driving, another was college, another was having a career, another was having a husband, and the most important one of all was being a mother," she says.

By the time she was in her early 20s, Wiparina had gone to college and learned to drive a van with joy stick computer control. She studied to be a school teacher, but had to abandon that career after contracting viral pneumonia from her students and spending three months in the hospital. "The doctor told me, 'If you want to live, you better not teach,'" Wiparina said. She got a job in marketing instead.

Wiparina married her college sweetheart, a tire store manager, and the couple began to investigate the idea of having children. "I'd always been told, 'You should never try to have kids, there's no way,'" Wiparina said. "We were scared to death because we didn't know anything."

Wiparina contacted a high-risk obstetrician, David Colombo, at Ohio State University Medical Center in Columbus, who agreed to help her. He assembled a team, including an anesthesiologist and another high-risk obstetrician who had experience doing Caesarean deliveries under local anesthesia in Africa.

Wiparina had no problems conceiving, and her two pregnan-



Stacy Wiparina and her baby on the day of the child's baptism

cies went fine, other than the babies growing to the left side of her belly, rather than to the front (a result of her scoliosis, or spinal curvature).

The births, unlike the pregnancies, required special consideration. Because of the spinal fusion Wiparina received at age 12 — "Otherwise I would have to lie down my whole life" — she could not get epidural anesthesia. Moreover, general anesthesia would be risky for a person with SMA.

Wiparina never went into labor, although it isn't clear that her SMA had anything to do with that. She lacked the strength to push the baby out, and in any case, her pelvis was too small for the baby to pass through. When the time came, the doctors performed a Caesarean (C-section) using several dozen shots of local anesthetic as the only pain relief.

"I felt the first few shots," Wiparina reports. "I didn't look. Once they started to cut, I didn't feel a whole lot, except for a really horrible sensation when they were putting my insides back in place." The experience was frightening, Wiparina said, especially since she was wide awake the whole time.

Wiparina went through an identical procedure for the birth of her second child, but "it wasn't as bad, because I knew what was coming." Besides, she adds, lots of women have to go through more than 45 minutes of discomfort in order to give birth.

Since the birth of her second child, Wiparina has developed symptoms of a compressed cauda equina (a bundle of nerves at the base of the spine that looks like a horse's tail). The cauda equina can become compressed in healthy women, but in Wiparina's case, carrying two babies to term while sitting in a wheelchair almost certainly caused the condition.

Wiparina has now lost most sensation below her waist, and it's doubtful it will ever return. Surgery might be able to correct the condition, but it's too risky.

Wiparina doesn't regret having children, though. She achieved her fondest dream and greatest goal, and if that's the price she has to pay for it, she says, so be it.

continued from page 6...

correct them (spinal fusions, often involving insertion of instruments, such as rods and screws) — can be a problem.

The growing uterus can press on an abnormally shaped or fused spine, possibly causing loss of sensation, increased weakness or even paralysis. (See "Achieving All Her Goals," above.)

Because of increased pressure and

strain on the lower back during pregnancy, compression of the cauda equina (the "horse's tail" formation of spinal nerves in the lumbar and sacral areas) does occasionally occur, even in healthy women, says Hannah Briemberg. The risk may be greater in neuromuscular diseases, such as spinal muscular atrophy. However, notes Briemberg, "It's still an extremely rare complication." She cautions that low back pain affects

almost everyone during pregnancy, and the vast majority of the time, it does not mean anything is seriously wrong.

The powers

The first stage of labor is one in which the cervix (the neck of the uterus) dilates so that the baby has room to leave the uterus during the second stage, when the baby is pushed out.

The first stage is entirely the result

of smooth muscle uterine contractions, which are involuntary. Women with **neuromuscular diseases** (with the exception of **myotonic dystrophy**) generally don't have any special problems with this stage of labor.

However, almost all women with neuromuscular diseases, (including those with myotonic dystrophy) have voluntary muscle weakness and have some difficulties with the second stage of labor.

The uterus continues to contract during the second stage, helping to

expel the baby. However, the mother's voluntary efforts, by contracting her abdominal muscles, add force in pushing the baby out in a timely manner.

"It's more difficult to push when the pushing stage comes, if you don't have much strength to begin with," says Briemberg. "In women who have prolonged labors, having some voluntary muscle strength to push could help."

That doesn't necessarily mean women with neuromuscular disease have prolonged or difficult labors or that operative interventions will be



Stacy Wiparina's uterus expanded to the side because of her spinal curvature.

Worth the Effort

Vickie Jahaske, 46
Tucson, Ariz.
dermatomyositis

Vickie Jahaske had always dreamed of having children, but that dream almost died with her diagnosis of **dermatomyositis** at age 23.



Vickie Jahaske and her second baby

Her symptoms began with weakness and pain in her legs and arms, and a rash on her face that she thought was caused by allergies. She had just gotten married, and, she recalls, "I tried to ignore my symptoms until I was unable to hold up my toothbrush."

Unlike many with this disease, Jahaske received a correct diagnosis right away, and began treatment with the anti-inflammatory medication prednisone and the immunosuppressant methotrexate. Even so, her symptoms worsened to the point where she needed a feeding tube to eat. "I remember asking the therapist, 'Will I ever swallow again?,' and she didn't know the answer," she says.

Jahaske fell into a deep depression that was only alleviated when her prednisone dose was reduced. She recovered somewhat and went back to work, in the legal department at Motorola.

"I made an agreement with my doctor that if I could stay off the methotrexate for six months, he would allow me to get pregnant."

Jahaske's first child, a healthy boy, was born when she

was 27. The pregnancy was normal — in fact her rash even improved — but when her water broke and labor failed to progress, she had to have an emergency Caesarean. After the birth, her dermatomyositis flared again. She went back on methotrexate and also began receiving intravenous infusions of immunoglobulins (another immunosuppressive treatment) every three months for years. "I never regained the amount of health I had before the pregnancy," she says.

Her health declined even further during her second pregnancy, when she was 33. "This one was difficult. My doctor told me I was giving him gray hair."

Jahaske developed high blood pressure, diabetes, a deficiency of blood-clotting cells and hair loss, all probably related to her anti-inflammatory and immunosuppressant medications. She also developed calcinosis, hard lumps of calcium under her skin, caused by her dermatomyositis, and the disease-related rash on her back and face spread.

Jahaske gave birth vaginally to a healthy girl, but within days, she began to lose her eyesight. Her vision grew so blurry and dark she could hardly see the baby while nursing. Her high blood pressure had caused macular edema, or swelling in her eyes. "Once again, the doctors could not predict if I would recover or not."

Jahaske slowly recovered, and her vision returned to normal, but she never went back to work after her second child. Her energy level dropped, and she continues to have high blood pressure, diabetes and the painful symptoms of dermatomyositis. The red rash on her back is as big and hard as a dinner plate. She wears caps, bandannas, and wigs to cover the rash and her few wisps of hair.

Although her pregnancies apparently worsened her fragile health, Jahaske said she is grateful to be able to have children, for her husband's unwavering support, and for the help she's received from MDA.

needed, but it does put women with these disorders at increased risk for needing them.

“As far as the actual labor and delivery are concerned, most of the force is generated by the uterus, which is smooth muscle,” Pressman says. “The uterus is quite capable of expelling a baby with no effort at all on the mother’s part. So, even though we encourage women to push, they don’t always need to push. Sometimes that additional effort is helpful, but much of the time, it’s not needed.” However, she notes, if “it’s a very snug fit,” or if the uterus itself is not functioning properly, physicians may intervene.

Pain management

Spinal abnormalities or the effects of spinal surgery can make regional anesthesia, such as spinal or epidural blocks, difficult. These types of anesthesia may not work as well in patients with significant spinal abnormalities or who have had instruments such as metal rods inserted.

“It’s not necessarily contraindicated to get these types of anesthesia,

but they may not work as well,” says Pressman. “Any time you’ve operated on the spine, there can be scarring, and so getting the medication to spread properly is difficult. If the space between the spinal elements is narrowed because of a change in diameter of the spine, then it can be hard to get the anesthetic in as well.”

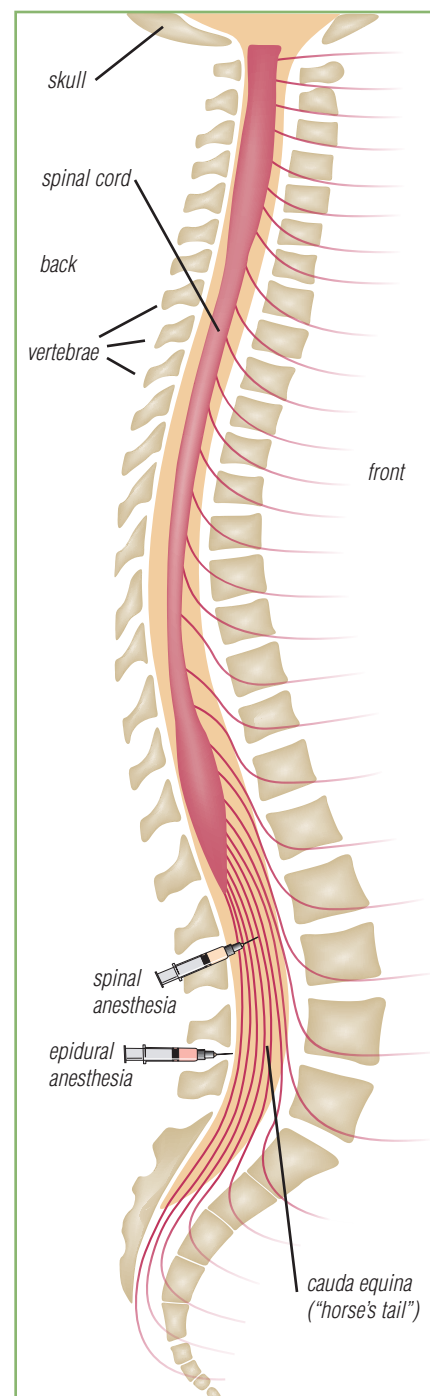
The same barriers apply to spinal or epidural anesthesia. The difference in these is in the exact placement of the anesthesia-delivering catheter (see illustration, at right).

On the other hand, general anesthesia (inhaled anesthesia), which is an alternative to epidural or spinal anesthesia for C-section deliveries, poses other hazards for some women with neuromuscular disease.

Women with central core disease, for example, are at very high risk for a severe adverse reaction called “malignant hyperthermia,” when given certain inhaled anesthetics. It’s very important that the obstetric team know about this risk beforehand, so they can use alternative anesthetic agents.

And, once again, the woman with

The Spine and Spinal Cord



Spinal curvatures and the spinal fusion procedures sometimes used to treat them can interfere with the effectiveness of spinal or epidural anesthesia. The medication-delivering catheter for spinal anesthesia is usually placed a little higher and penetrates a little more deeply than the catheter to deliver epidural anesthesia.

Good Advice from Women Who’ve Been There

Women with neuromuscular disease who’ve gone through pregnancy — and the medical experts who care for them — offer this advice:

- Choose a physician who is knowledgeable, prepared and enthusiastic about helping you have a child. If your doctor is unsympathetic or unsupportive, find another one.
- Ensure communication among all members of your medical team — neurologist, obstetrician, anesthesiologist, primary care doctor, pediatrician, etc. All must be aware of your health status and prepared for the possibility of special complications.
- As far in advance of the big day as possible, decide with your doctor where and how you will give birth. Make sure the hospital has a neonatal intensive care unit that can handle potential complications.
- Arrange for help once you come home, to avoid overwork and injury. One way to easily mobilize a network of family and friends is [myMuscleTeam](http://www.mda.org/mymuscleteam) (see www.mda.org/mymuscleteam), MDA’s care coordination website, which can help you (or a caregiver) recruit and schedule willing volunteers.

The Nurse Called it 'Fred's ataxia'

Beth Bax, 41
Altadena, Calif.
Friedreich's ataxia

Like many young mothers with (and without) neuromuscular disease, Beth Bax discovered that pregnancy and childbirth can be a piece of cake compared to caring for a newborn.

Bax, a 41-year-old engineer, received a diagnosis of **Friedreich's ataxia (FA)** in December 1999, at age 30. She'd been married two months before. Her speech was slurred and something was "off" with her balance — "I looked a little drunk probably," she says — but she was not yet using a cane.

Bax gave birth to her first child in December 2001. The pregnancy was uneventful; her obstetrician researched FA and didn't think it would affect the birth.

Bax had epidural anesthesia, and the birth went fine, other than the inappropriate behavior of the attending nurse. "I mentioned to her that I had FA, and she freaked out and ordered a baby warmer and extra nurses," Bax said. "I didn't want to calmly explain that I was not giving birth to the elephant baby while the nurse was telling everyone I had *Fred's ataxia*. She never did get the name right."

While FA didn't have a big effect on Bax's pregnancy or birth experience, it definitely affected her confidence as a new mother. "My husband ended up always carrying her because I needed my hands to grab the walls," Bax says. "I played with her on beds and couches and didn't carry her much at all."



Beth Bax and her daughter

By the time of her second pregnancy, in the summer and fall of 2004, Bax had begun using a walking stick. ("They're so much cooler than canes," she reports.) She had extra visits with her neurologist at the University of California-Los Angeles, was able to exercise, and maintained good circulation and stable health. Doctors induced the birth, Bax had epidural anesthesia, and daughter Charlotte came out fine.

FA again complicated matters after the second baby was born, Bax says. "I did not carry her at all. I put her in a stroller just to move from room to room at my house. When she was 3-and-a-half months old, I was leaning down to pick up a baby sock off the floor, and I fell."

Bax couldn't get up. She'd broken her leg and torn her knee ligament, necessitating wearing a long leg cast for several months. "It was very difficult," she says. "My FA made my balance terrible, and now I had to balance on one leg." She couldn't drive, get in the shower without help, or walk down the steps to leave the house. She began using a wheelchair full time.

"Eventually, everything was fine but it took a while. We got a full-time nanny, we ramped the steps, and I learned to carry the baby on my lap as I moved forward with my free hand and my one good leg. We made adjustments and survived, but I never went back to walking. The muscles for standing and walking had atrophied, and I never wanted to be in a position where I might break a leg and be helpless again."

Bax believes that having children probably sped up the course of her FA, but she has no regrets. Rather than thinking about symptoms and disease progression, "I spend a lot of time thinking about laundry, what we can eat for dinner, getting homework done, getting the house clean, which school my kids should go to, etc. I don't have time to think about FA a lot. If I focused on all the problems I have, I think my FA symptoms would be more pronounced and I would be more depressed about it."

myotonic dystrophy is at high risk of an adverse reaction (though not necessarily malignant hyperthermia) to general anesthesia and, in her case, also to the pain-killing medications that are sometimes given intravenously during labor.

"The big concern is for the anesthesiologist and obstetrician to be aware that they might be a little bit more sensitive to these intravenous medications that are given during labor and

maybe to not dose quite as heavily as they would in a woman who didn't have myotonic dystrophy," Briemberg says.

If a woman needs general anesthesia during a surgical delivery, she's generally "intubated," meaning the surgical team puts a tube down her trachea, and a ventilator breathes for her during the surgery. In the average person, the tube can be removed almost immediately after the delivery, and she can breathe

on her own without difficulty.

However, women with very weak respiratory muscles, such as someone with spinal muscular atrophy or uncontrolled myasthenia gravis, may have great difficulty resuming normal respirations after having been intubated, even for a short time.

Fearing this complication, some women have opted for an unusual way to manage pain relief during a C-section



Having a neuromuscular disease can pose special anesthesia-related risks.

— local anesthesia. (See [“Achieving All Her Goals,”](#) page 9.)

David Colombo, who delivered Stacy Wiparina’s babies by C-section, used local anesthesia both times. “We have a whole protocol that involves 100 cc of lidocaine,” Colombo says. “I numb the skin, and then we make our incision. Then you go into the fascia [layer of tissue just under the skin], and you incise that, and then you numb the peritoneum [deeper layer of tissue] over the uterus. They don’t feel it when you cut into the uterus; there’s no sensation there. Actually, people tolerate it very well. If you take your time and do it right, it’s just the same as a spinal. Nobody really complains. And you don’t have to worry about intubation.”

Eva Pressman, however, isn’t as sure about using local anesthesia for a C-section. “I’ve seen it done a couple of times,” she says. “You can numb the skin and the fascial layers very effectively, but you can’t numb the peritoneum very well. Once you get into the abdominal cavity, you get pretty uncomfortable. That being said, I worked with an older obstetrician 20 years ago who had three C-sections herself, all under local anesthesia, because that’s what she wanted. It would not be my recommendation, but if you get the right sort of person, you can do some things that other people

might not be able to tolerate.”

Pregnancy and disease progression

“I think the big thing is that, almost as a rule, you have to expect that you’re going to get a little bit weaker during the pregnancy and, unfortu-

nately, that you’re probably not going to bounce back after the delivery,” Briemberg says. “Why they get weaker, I don’t think we know. It doesn’t happen to everyone, but it certainly does happen to a significant number, so I think they have to be prepared for that.”

Even if weakness doesn’t increase permanently, the pregnant woman with a neuromuscular disease is almost certain to experience a temporary ratcheting up of her disability level, says Pressman. “Pregnancy is a time in your life when you’re gaining more weight than you would normally gain in a short period of time,” she notes. “So, if you have enough muscle strength

to ambulate and carry on your routine activities, but you gain 20 or 30 percent more weight over the course of the pregnancy, you might not be able to continue those sorts of things. You also change your center of gravity, so you use different muscles to support and balance yourself, and your weakness might become more evident.”

Teamwork makes the difference

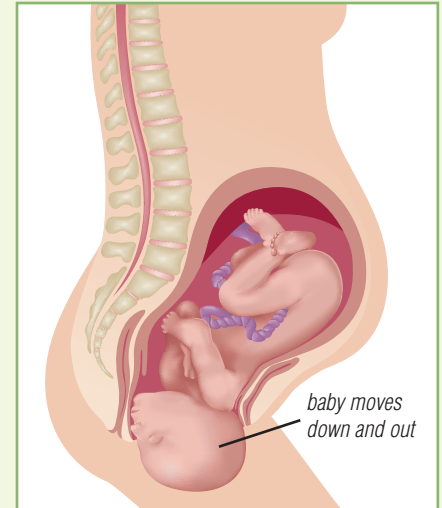
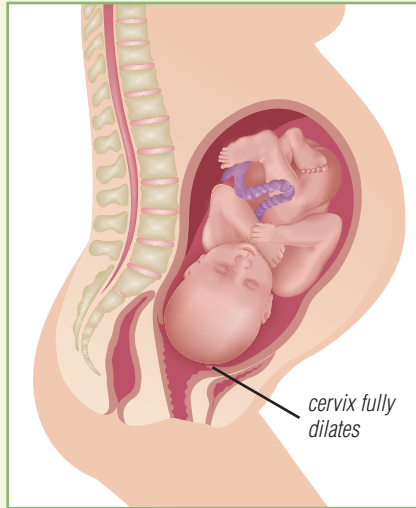
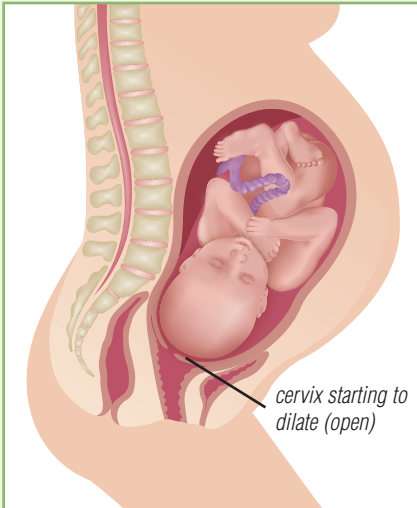
Coordination and communication among the pregnant woman, her neurologist and a team experienced in high-risk obstetrics (such as can be found at most major medical centers) are essential to a good outcome, experts agree. In addition, meeting with a genetic counselor can help couples assess the risk of passing a genetic disorder to a child. Carrier testing and prenatal diagnosis are available for many diseases. (See [“The Pain and Promise of Prenatal & Newborn Genetic Diagnosis”](#) in the [July 2007 Quest](#).)

“Care that is well coordinated among services is safe care, and care that is done in isolation can be less safe,” Pressman says. “It’s really important that the different care providers communicate with one another and



Coordination among members of the health care team makes care safer.

Stages of Labor



During the first stage of labor, the cervix (neck of the uterus) dilates. This stage involves the uterine muscle, which is involuntary. This type of muscle, also known as "smooth" muscle, has normal function in most neuromuscular diseases. An important exception is myotonic dystrophy, in which uterine and other smooth muscle function can be impaired.

During the second stage of labor, the uterus continues to contract, but it's now aided by the mother's voluntary muscle efforts to push the baby out. Voluntary muscles in the abdomen can be weak in neuromuscular disease, interfering with the expulsion of the baby and sometimes leading to an assisted vaginal or a Caesarean delivery.

that the patient tells the same things to different providers. I've had patients who have come to me and who've sort of downplayed their illness. Then I get records from their neurologist, and it turns out they've had many more complications than they've been willing to share with me."

Pressman recommends that, ideally, women with a **neuromuscular disorder** have a consultation with a physician with expertise in complicated pregnancies before becoming pregnant. "Once you're pregnant," she notes, "there are fewer options."

Briemberg adds, "It's important to have a pre-delivery anesthetic consult, in case you need general anesthesia." If anything, she says, women with neuromuscular diseases "end up being safer than women that don't have a neuromuscular disease," if they have a preoperative anesthetic consult, because "everybody's very in tune to all the issues and knows what agents can and can't be used."

Worth it

"My experience is that most people feel it was still worthwhile having the pregnancy," says Briemberg. In fact, she says, it's important for doctors to "be realistic," while at the same time not causing women to worry too much.

"We're all so anxious when we're pregnant, about everything," she says, "and I really try more than anything to be reassuring. For most of these women, it's not going to have a huge impact on their disease. They're going to have all the trials and tribulations of being a parent, but for the most part, the disease isn't a huge additional factor." □



Beth Bax says having children probably sped up the course of her disease, but she has no regrets.

A Turn of Fate

**Erin Brady Worsham, 51
Nashville, Tenn.
amyotrophic lateral
sclerosis (ALS)**

Erin Brady Worsham tried to get pregnant for six years, with no luck. She had accepted that she was never going to be a mother. Then, in a turn of fate worthy of a novel, she got pregnant the day after she received a diagnosis of **amyotrophic lateral sclerosis (ALS)**.

Worsham was 36 at the time. She had been having trouble with foot drop (an inability to lift the front of the foot) and weakness in her lower legs, so she went to a neurologist who did a series of tests. On Sept. 7, 1994, Worsham and her husband met with the doctor for the verdict.

“His nurse put her arm around my shoulders and walked me to his office,” Worsham recalls. “I knew I was in big trouble.”

When she learned of her pregnancy, Worsham was delighted. Her neurologist, however, was not, commenting, “Don’t you know you could be gone in a year?” Worsham went in search of a doctor who would be more supportive, and found Nashville obstetrician John Vanhooydonk.

“I remember Dr. Van saying, ‘It doesn’t matter if you live five days, five months or five years, you’re going to be happy you had this child.’”

Worsham felt great throughout her pregnancy: “Food tasted incredible. I was not alone. My thoughts were of life, not death.”



But ALS began to take its toll. Worsham’s leg strength and balance were compromised, and worsened as the baby got heavier. She began wearing shoes to support her ankles and had braces made, which she still wears, to address her foot drop. She started walking with a cane.

She also had two or three instances of choking. “The first time it happened was very scary, because it was so unexpected,” she says. “Instead of being able to just clear my throat, my throat completely closed up and I couldn’t breathe in or out at all. At first I panicked, but then, instinctively, I forced myself to relax and my throat relaxed too. From then on, I was very conscious of my chewing and swallowing.”

For the birth, Worsham arranged for an anesthesiologist to give her an epidural, but she didn’t need it. She described the natural childbirth of her son Daniel as follows:

“Dr. Van saw on the ultrasound that Daniel was turned face up. He warned us that I would be facing a C-section [Caesarean delivery] if the baby didn’t turn. My nurse friend and [my husband] Curry turned me as far as they could on my stomach. They gave me a “whiff” of stadol [a narcotic painkiller]. It didn’t take away the pain, but it allowed me to rest between contractions. A doctor friend of ours dropped by to see how I was doing. She made the mistake of holding my hand. I never let her go.

“Dr. Van returned when I was fully dilated. [The cervix, or entrance to the uterus, must be fully dilated before the baby can pass through.] The ultrasound confirmed Daniel had indeed turned. Dr. Van commented that there was ‘a lot of power’ in the room. With my nurse friend pushing on one leg and Curry pushing on the other and my doctor friend pushing on my back, we brought Daniel home.”

Now 51, Worsham has outlived all the doctors’ predictions, and Daniel, at 15, is a strapping young man. Worsham advises women with ALS who want children, “Don’t wait! The earlier in the disease, the better.”

Surprise Pregnancies

Jeanne Lawrence, 34
Quincy, Ill.
dermatomyositis

After all the treatments she'd undergone since learning at age 14 that she has **dermatomyositis (DM)**, Jeanne Lawrence was sure she'd never have kids. She'd taken large doses of prednisone, immunoglobulins, and Imuran (azathioprine), among other things. That's why she was surprised and scared to learn, shortly after marrying at age 25, that she was pregnant. At the time, she also was undergoing plasmapheresis, a treatment that filters unwanted antibodies from the blood that's sometimes used to treat autoimmune diseases like DM.

"I didn't know if the plasmapheresis would hurt the baby," she recalls. "But the doctor read up on it and said, 'Go ahead.'"

Lawrence had high blood sugar and pre-term labor, but was able to give birth vaginally. Her first son came into the world in 2001, five-and-a-half weeks premature and with pneumonia, but is healthy now. When Lawrence took him in for his six-week checkup, she was shocked to learn she was pregnant again.

The second pregnancy went much like the first. Her doc-



tor put her on blood sugar medicine, which made her so light-headed she couldn't walk. She went through another plasmapheresis treatment while pregnant.

Ten months and one week after having her first child, Lawrence had her second, a healthy girl. She again gave birth vaginally, without complications, although the baby weighed almost nine-and-a-half pounds. "The doctor looked at me and said, 'You are planning on waiting this time [before getting pregnant], aren't you?'"

Lawrence had a miscarriage in 2005, then underwent her third and final pregnancy later that year. "I was miserable," she says. She couldn't sleep, experienced some spotting, and her hormone and blood sugar levels were way out of whack. "I had to go to the hospital in Springfield for ultrasounds every two weeks, then every week. At 36 weeks, I measured like 50 weeks. The doctors were worried I had so much fluid that I wouldn't be able to deliver the placenta."

Lawrence's last child, a boy, was born in 2006 via Caesarean section. He weighed more than 11 pounds and required treatments prior to delivery to help his lungs mature, but otherwise was healthy.

Lawrence says it's difficult to care for her children. She suffers flare-ups of dermatomyositis and tires easily. "I sometimes have to crawl up or slide down stairs, and I can't stand extremes of temperature. I have to stay inside a lot."

She encourages women with muscle disease who are contemplating pregnancy to make sure they have good family support. "My mom was there for me through every pregnancy," says Lawrence, who is now divorced. "I am very blessed to have both her and my children in my life."

Medication Complications for Pregnant Women with Neuromuscular Disease

Medication or Treatment	Prescribed For	Potential Problems During Pregnancy
alglucosidase alfa (Myozyme)	acid maltase deficiency (Pompe disease)	<ul style="list-style-type: none"> No studies have been conducted with this drug in pregnant women. In short-term studies in a mouse model, this drug did not impair fertility or harm a growing fetus. No long-term studies in animals have been performed.
azathioprine (Azasan, Imuran)	dermatomyositis inclusion-body myositis polymyositis myasthenia gravis	<ul style="list-style-type: none"> Can affect fertility in both men and women. Animal studies have revealed evidence that use of azathioprine can cause malformations in a fetus. Has been shown to cross the placenta and cause harm to the fetus, with some data showing an increased risk of intrauterine growth retardation, premature birth and low birth weights. Anecdotal evidence exists from cases in which the drug was used during pregnancy by women with conditions such as kidney transplant or systemic lupus. Congenital anomalies were observed in children born to these women, including: missing or extra fingers or toes; congenital heart disease; umbilical hernia; deformities in the legs, feet, penis and skull. Some babies had problems producing bone marrow.
baclofen (Kemstro, Lioresal)	amyotrophic lateral sclerosis	<ul style="list-style-type: none"> No adequate and well-controlled studies in pregnant women have been conducted. Female rats treated with baclofen experienced an increase in incidence of ovarian cysts. Animal studies in rats have revealed an increased incidence of birth defects. Abnormalities in the bones of the forelimbs and hindlimbs were observed in fetuses of rabbits treated with the drug.
clonazepam (Klonopin)	amyotrophic lateral sclerosis	<ul style="list-style-type: none"> Limited experience with the use of clonazepam during pregnancy has been reported, but an increased risk of birth defects has been associated with use of all known anticonvulsant agents, such as clonazepam. Breathing and feeding problems in newborns have been observed.
corticosteroids prednisone tablets (Deltasone) intravenous methylprednisolone sodium succinate (Solu-Medrol)	congenital MD distal MD Duchenne MD Emery-Dreifuss MD facioscapulohumeral MD limb-girdle MD myotonic MD oculopharyngeal MD myasthenia gravis	<ul style="list-style-type: none"> Studies in animals have shown that corticosteroids cause birth defects. Studies on correlation of birth defects with corticosteroids have not been done in humans. Anecdotal evidence reported by medical professionals suggests a lack of abnormalities in children whose mothers were treated with typical doses of prednisone and methylprednisolone throughout pregnancy. Clinical experience has revealed that corticosteroid use in pregnant women may correlate with premature rupture of amniotic membranes and low birth weights in infants.

Medication Complications for Pregnant Women with Neuromuscular Disease

Medication or Treatment	Prescribed For	Potential Problems During Pregnancy
cyclophosphamide (Cytoxan, Cytoxan Lyophilized, Neosar)	dermatomyositis inclusion-body myositis polymyositis	<ul style="list-style-type: none"> • May affect egg production in women and sperm production in men. Use of cyclophosphamide by the father prior to conception has been associated with birth defects. • Normal pregnancies and fetal development have been observed in women who used this drug during pregnancy, but it also has resulted in severe and multiple birth defects in some instances.
cyclosporine (Gengraf, Neoral, Sandimmune)	dermatomyositis inclusion-body myositis polymyositis	<ul style="list-style-type: none"> • Human data have revealed evidence of premature birth and low birth weight in babies born to women who took this drug during pregnancy, but the drug does not appear to cause birth defects. • In data gleaned from 15 studies, analysis showed that the overall prevalence of major birth defects in babies born to women taking this drug did not vary substantially from that reported in the general population.
dantrolene (Dantrium)	Used preoperatively to prevent or halt the development of malignant hyperthermia	<ul style="list-style-type: none"> • The safety of this drug in women during or prior to pregnancy has not been established.
entanercept (Enbrel)	dermatomyositis inclusion-body myositis polymyositis	<ul style="list-style-type: none"> • Data from human pregnancy studies are not available. • Developmental toxicity studies performed in rats and rabbits at doses ranging from 60 to 100-fold higher than the human dose have revealed no evidence of harm to the fetus.
gabapentin (Gabarone, Neurontin)	amyotrophic lateral sclerosis	<ul style="list-style-type: none"> • It is not known whether Neurontin is harmful to an unborn baby, but some observations have indicated that gabapentin does cross the placenta and accumulate in the fetus. • In animal studies, the drug proved toxic to the fetus, causing delayed formation of several bones and accumulation of urine in the kidneys.
hydroxychloroquine sulfate (Plaquenil, Quineprox)	dermatomyositis inclusion-body myositis polymyositis	<ul style="list-style-type: none"> • There are no controlled data in human pregnancy. However, the findings of one study have shown preliminary safety support for the use of this drug during pregnancy. • Animal studies have revealed that the drug passes rapidly across the placenta, accumulates selectively in the eyes of the fetus and still is present in those tissues five months after elimination from the rest of the body.
infusion of mixed immunoglobulins (Gammar, Gammagard, Sandoglobulin, others)	dermatomyositis inclusion-body myositis polymyositis myasthenia gravis	<ul style="list-style-type: none"> • IVIg has been shown to cross the placenta after 32 weeks gestation, but successful outcomes in human pregnancy have been observed. • Noted risks for the use of IVIg in pregnancy are blockage of a blood vessel by a blood clot, and kidney inflammation • Animal studies have not been reported.

Medication Complications for Pregnant Women with Neuromuscular Disease

Medication or Treatment	Prescribed For	Potential Problems During Pregnancy
magnesium	amyotrophic lateral sclerosis	<ul style="list-style-type: none"> • Studies in pregnant women who were treated with magnesium sulfate by injection showed no risk of increased fetal abnormalities. • Magnesium sulfate often is used to treat severe high blood pressure during pregnancy. • It should not be prescribed to pregnant women with myasthenia gravis, as it is known to exacerbate the disease. • Although the drug has a depressant effect on the central nervous system, evidence has shown it does not adversely affect the mother, fetus or newborn when used to treat high blood pressure and fluid retention (pre-eclampsia) or the convulsions or coma that may follow (eclampsia).
methotrexate (Rheumatrex, Folex, Mexate)	dermatomyositis inclusion-body myositis polymyositis	<ul style="list-style-type: none"> • May cause birth defects and fetal and newborn death if taken during pregnancy. • Pregnancy should be avoided if either partner is receiving this drug. Men should wait three months after therapy, and women should wait at least one ovulatory cycle before trying to conceive a child.
mycophenolate mofetil (CellCept)	dermatomyositis inclusion-body myositis polymyositis myasthenia gravis	<ul style="list-style-type: none"> • There are no controlled data in human pregnancy, but the drug has been associated with increased risk of first trimester pregnancy loss and increased risk of birth defects. • National Transplantation Pregnancy Registry (NTPR) data on mycophenolate mofetil-exposed pregnancies in transplant patients showed that 45 percent of pregnancies ended in spontaneous abortions. Of live-born babies, 22 percent had birth defects.
nabumetone (Relafen)		<ul style="list-style-type: none"> • There are no controlled data in human pregnancy. • Animal studies have not shown this drug to cause birth defects, but there has been evidence both in animals and humans of spontaneous abortion. • It's advised that this drug not be used in the third trimester, as it may affect the fetus' cardiovascular system, and also prolong labor and delivery.
neostigmine (Prostigmin, Prostigmin Bromide)	myasthenia gravis	<ul style="list-style-type: none"> • There are no adequate studies of the use of this drug in pregnant women.
Plasmapheresis Note: plasmapheresis is a procedure, not a medication	dermatomyositis inclusion-body myositis polymyositis myasthenia gravis Lambert-Eaton syndrome	<ul style="list-style-type: none"> • May alter blood volume in the mother and cause low blood pressure, which can endanger both mother and fetus.

Medication Complications for Pregnant Women with Neuromuscular Disease

Medication or Treatment	Prescribed For	Potential Problems During Pregnancy
pregabalin (Lyrica)	amyotrophic lateral sclerosis	<ul style="list-style-type: none"> • There are no controlled data on the use of this drug in human pregnancy. • There is evidence that men being treated with this drug have an increased risk of contributing to birth defects in offspring. • Animal studies have shown that pregabalin crosses the placenta and has resulted in increased incidences of malformations, slowed growth, nervous and reproduction system impairment, decreased fetal body weights and fetal death.
pyridostigmine bromide (Mestinon)	myasthenia gravis	<ul style="list-style-type: none"> • No information on the safety of this drug during pregnancy in humans has been established. • Drugs that are biochemically similar to pyridostigmine bromide have not been reported to cause birth defects; however, temporary muscle weakness has occurred in some newborns whose mothers took such drugs during pregnancy.
Quinine (Qualaquin)	amyotrophic lateral sclerosis	<ul style="list-style-type: none"> • There are no controlled data on use of this drug in human pregnancy. • Animal studies have revealed evidence that the drug causes fetal malformations. • It's known that quinine crosses the placenta and accumulates in fetal blood. • In a study of women with a form of malaria, birth defects were observed in 21 infants exposed to high doses of quinine during the first trimester.
riluzole (Rilutek)	amyotrophic lateral sclerosis	<ul style="list-style-type: none"> • There are no adequate and well-controlled studies in pregnant women. • At higher-than-normal doses, riluzole impaired fertility when administered to male and female rats. • In studies in rats given the drug prior to and during mating and pregnancy, riluzole caused decreased implantations, increased intrauterine death and viability and growth of offspring. • Administration of higher-than-normal doses of the drug to pregnant rats and rabbits proved toxic to both fetus and mother.

Medication Complications for Pregnant Women with Neuromuscular Disease

Medication or Treatment	Prescribed For	Potential Problems During Pregnancy
<p>succinylcholine (Anectine)</p> <p>Note: This muscle relaxant is a “depolarizing” relaxant often used with anesthesia.</p>	<p>May be of particular concern in:</p> <p>amyotrophic lateral sclerosis central core disease myasthenia gravis myotonia congenita myotonic muscular dystrophy and some other MDs periodic paralysis spinal muscular atrophy</p>	<ul style="list-style-type: none"> • Can trigger a malignant hyperthermia reaction in people with central core disease, myasthenia gravis and some muscular dystrophies. (Malignant hyperthermia is a potentially fatal abnormal response to inhaled anesthetics and certain muscle relaxants. It causes uncontrolled muscle contractions, accelerated metabolism, high fever, and, if not treated, death.) • Can affect heart rhythm in people with spinal muscular atrophy or ALS. • Can cause prolonged weakness after surgery in those with periodic paralysis. • Can make muscles rigid, complicating some medical procedures, in people with myotonic muscular dystrophy or myotonia congenita. • It is not known whether succinylcholine adversely affects reproductive capacity, or if it causes fetal harm when administered to a pregnant woman. No information on the safety of this drug during pregnancy in humans has been established. • Animal reproduction studies have not been conducted with succinylcholine chloride. • A higher number of women who are pregnant, versus those who are not, may have increased sensitivity to the drug (prolonged duration of slowing of breathing). • Small amounts of the drug are known to cross the placenta. Depending on the dose given the mother during delivery, a newborn may experience slowed breathing and lack of muscle tone.
<p>tacrolimus (Prograf)</p>	<p>dermatomyositis inclusion-body myositis polymyositis</p>	<ul style="list-style-type: none"> • In a study of 100 pregnancies in 84 mothers, there were 68 live births in which the most common complications to the newborns were temporary low oxygen levels, raised potassium levels in the blood and kidney problems. • Some animal studies have revealed an increased incidence of abortion, resorption of the fetus, and malformations. The drug also was found to be toxic to the mother. • It's known that Tacrolimus crosses the placenta. • The drug has been used successfully during pregnancy in a limited number of cases, although raised serum potassium levels and kidney problems in newborns have been reported.
<p>tizanidine (Zanaflex)</p>	<p>amyotrophic lateral sclerosis</p>	<ul style="list-style-type: none"> • There are no controlled data on use of this drug in human pregnancy. • Animal studies have shown evidence of increased length of gestation, increased loss of pups, and impaired development.
<p>ubiquinone (Co-Q10, Coenzyme Q10, Idebenone, LiQsorb, Liquid Co-Q10, NutraDrops, QuinZyme, others)</p>	<p>Friedreich's ataxia</p>	<ul style="list-style-type: none"> • Information regarding the safety of ubiquinone in pregnancy is lacking, but its use is not recommended. • Animal studies have revealed no harmful effects.