

Treatment can allow birth despite dangerous disorder

October 8 2009, By John Fauber

It's a decision that an expecting mother should never have to make: Abort your unborn child and save your own life or deliver the baby and face possible death a few days later.

Those were the awful choices facing Terrilyn Priessnitz a few weeks after she became pregnant with her first child earlier this year.

Priessnitz, 36, developed shortness of breath in March and a short time later was told she had pulmonary hypertension, an often fatal lung disorder.

Along with the diagnosis came a terrible statistic: Women with pulmonary hypertension who give birth face up to a 50 percent chance of dying shortly after the baby is born. The usual recommendation is to abort the pregnancy.

"It was hard, the thought of ending it," Priessnitz said from her bed at Aurora Sinai Medical Center, a few days before she gave birth last week. "Either I have this baby or there are no babies."

Medical literature is replete with warnings about the combination of pulmonary hypertension and pregnancy.

The combination is so dangerous that women with the condition are advised to be sterilized.

Not found in the published annals of medicine is an [aggressive treatment](#) devised by a Milwaukee doctor that may allow women like Priessnitz to safely deliver their babies.

The protocol developed by cardiologist Dianne Zwicke, who practices at Aurora St. Luke's Medical Center, now has been used on 45 [pregnant women](#) with pulmonary hypertension, mostly in Milwaukee, all of whom survived along with their babies.

The treatment, which involves timing the delivery with the condition of the heart, could have worldwide implications for women with the disease. At least 100,000 people are known to have pulmonary hypertension in the U.S. and women are more than twice as likely as men to have it.

The key to the protocol is to constantly use ultrasound to monitor the condition of the right ventricle, the chamber of the heart that pumps blood into the lungs. Those echocardiograms are used to determine when to induce birth, although Zwicke does not allow the women to go beyond 36 weeks.

The women also receive IV diuretics to drain excess fluids, and are given other drugs. They are admitted to cardiac intensive care immediately after the baby is born and stay there for at least three days.

Doctors familiar with Zwicke's protocol warned that while it is promising, it is too preliminary to recommend it to women who are deciding whether to give birth.

"The risk is unacceptably high even if it (mortality) is only 10 percent," Uri Elkayam, a professor of cardiology and obstetrics and gynecology at the University of Southern California School of Medicine. "If you continue the pregnancy and deliver, you can die."

Elkayam did successfully use Zwicke's approach on a couple of patients, but cautioned that Zwicke's research is unpublished and has not been rigorously reviewed. In addition, it represents a series of case reports mostly from one hospital, not the experience of several doctors with large numbers of patients from a variety of institutions.

Pulmonary hypertension, which has several known causes but often develops for unknown reasons, is a condition in which the blood pressure is abnormally elevated in the pulmonary artery, the major vessel taking blood from the heart to the lungs. Priessnitz's pulmonary hypertension was caused by an autoimmune disease that formed blood clots in her lungs. While most of the larger clots have been dissolved, Priessnitz will be on blood thinners for the rest of her life.

The condition makes the heart work harder, a workload that becomes even greater because of the demands of pregnancy and giving birth.

Published reports place the maternal death rate at 30 percent to 56 percent, usually within 10 days of delivery.

Even for those who don't become pregnant the disease has no cure and the overall survival rate is poor, traditionally about three years from the onset of symptoms. However, several promising new treatments have become available in recent years and some people now can live with the disorder for 15 to 20 years. A lung or heart-lung transplant may be the only option for some patients.

Zwicke said that in addition to the women she has treated here, she also has consulted on several cases in other countries, all 45 with the same outcome: survival of the mother and baby. Still, she said, her approach should not be used to assure women that they can safely deliver their babies. At the same, she said she does not tell women they have to terminate their pregnancy.

"If you read the literature, it says you should abort," she said. "(But) what good am I to walk in and say, 'You have to abort?' "

Zwicke said she says tells women what the established research shows -- a 50 percent mortality rate -- and what her own clinical experience has been with the 45 women who survived.

"We don't push them either way," she said. "They are intelligent adults. It comes down to how much risk they are willing to take. I lose sleep and get gray hair over this."

Zwicke's research represents a promising start that someday may change clinical practice and allow doctors to reasonably assure women that the odds of dying are low, said Tracy Stevens, a cardiologist and professor of medicine at the University of Missouri -- Kansas City School of Medicine.

"Sadly, that will take years," she said.

One problem is that while the protocol used by Zwicke makes sense, it is very sophisticated and requires a variety of specialists. Many hospitals would not be able offer it, she said.

More importantly, the research needs to be duplicated in controlled clinical trials involving large numbers of women treated at different hospitals, she said.

It was only a year ago that Zwicke presented her findings at a national meeting of the American College of Chest Physicians. Hundreds of specialists were in the audience and many wrongfully thought she was saying that her approach could be safely applied to women all over the country, said Gerald O'Brien, a pulmonologist with the Christiana Care Health System in Wilmington, Del.

"She was attacked," O'Brien said. "A lot of specialists were saying you shouldn't even imply this."

About a year ago, Zwicke helped O'Brien use her protocol with a woman who learned she had [pulmonary hypertension](#) 30 weeks into her pregnancy. The mother and the baby both survived, he said.

Still, he said, he would be hesitant to try it again, at least for [women](#) who are in their first trimester.

"I would strongly encourage termination," O'Brien said. "Right now I don't think there is sufficient data out there."

Even knowing Zwicke's promising results, the decision for Priessnitz was difficult. Eighteen to 20 weeks into the pregnancy, she waffled between terminating it and continuing.

"I was a mess," said her husband, Daniel Bauer. "I didn't know what to think."

Priessnitz, of Racine, Wis., said she was encouraged by Zwicke's research so she continued with the pregnancy.

"It could have been that's what I wanted to hear," she said. "She (Zwicke) said I'll tell you if I think you should end this."

On Oct. 5, about 36 weeks into her pregnancy, Priessnitz was induced.

Finally, about 4 a.m. Oct. 6, a cesarean section was performed and 5-pound, 9-ounce Braden Bauer was born.

Zwicke said late last week that mother and child were doing well.

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