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THE LONG QT SYNDROME AND PREGNANCY - A.J. Morris MD, University of Rochester NY

The Long QT Syndrome (LQTS) is a genetically transmitted disorder that affects the electrical activity of the heart. Individuals affected with this condition may lead entirely normal lives, but others are at increased risk of developing serious heart rhythm disorders that may be manifest as passing-out spells (syncope) and/or sudden death. Fortunately, very good treatments are now available to stabilise the electrical circuits of the heart and to prevent dangerous changes in the heart beat.

For the past 15 years, the Rochester group, in conjunction with our medical colleagues at other academic institutions in the United States and Europe, have maintained an International LQTS Registry to better understand the natural history and clinical course of patients with this disorder in order to develop more effective preventive therapy. We have recently used this Registry that involves over 700 families with LQTS and more than 3,000 affected family members, to evaluate the effect of pregnancy on the occurrence of cardiac events in women with the hereditary Long QT Syndrome.

This pregnancy study was a retrospective analysis of 216 women with documented LQTS who had one or more pregnancies. The study population consisted of 111 probands (the first member of a family to be identified with LQTS, usually because of symptoms such as syncope) and 105 affected family members who generally had not experienced any symptoms. The incidence of heart events including syncope, aborted cardiac arrest, and sudden death was compared during equal 9 month pre-pregnancy, pregnancy, and post-pregnancy intervals in the probands and the affected family members.

What did we find. First, the 9 month interval during pregnancy and the delivery itself were not associated with an increased occurrence of heart events in any of the LQTS women. Secondly the 9 month period after pregnancy (the post-partum interval) was associated with an increased number of serious heart events in some women, especially the proband women who had experienced heart related symptoms (syncope) a year or more before the pregnancy. Thirdly, treatment with beta-blocker medication, standard therapy for symptomatic patients with LQTS, was associated with a significant reduction in the risk of serious heart events during and after the pregnancy. That is, LQTS women receiving beta-blocker medication did much better than those who did not receive this medication. Fourthly beta-blocker therapy had no adverse effects on the pregnancy or the foetus.

What can we conclude from this study? It appears that the physical and emotional stress during the post-partum period after pregnancy may be a factor in triggering heart rhythm disorders in some vulnerable women with LQTS. Certainly, the responsibilities of caring for a newborn, the interrupted sleeping schedule, the changing hormonal environment in the body, and fatigue, common features after pregnancy, may all be contributing factors to increase the occurrence of heart events during the post-partum interval. Every effort should be made to reduce the post-partum stress in the patients. Patients with LQTS who have had prior heart events should be prophylactically treated with beta-blocker medication, and this therapy is especially important in relationship to the pregnancy and should be continued during and after the pregnancy. Women with LQTS who are contemplating pregnancy should be seen in consultation by a cardiologist, and followed by the cardiologist during the course of the pregnancy, and afterwards.

We now have a better understanding that in LQTS women the risk of heart events increases in the 9 month interval after pregnancy. Furthermore, effective treatment is available in the form of betablockers to make pregnancy and the post-partum period safer. Clearly, an ounce of prevention is worth a pound of cure.