

Long QT Syndrome—Gender Associated Symptoms and Frequency of Cardiac Events During Pregnancy and the Post-Partum Period

Studies show there are significant gender differences in genotyped LQTS patients relative to age of first episode, syncope, cardiac arrest and sudden death. According to Emanuela H. Locati et.al. in the paper, “Age and Sex-Related Differences in Clinical Manifestations in Patients with Congenital Long-QT Syndrome”, males have a higher risk of cardiac events (meaning syncope or blackout spells, cardiac arrest and sudden death) during childhood (with an average earlier age of onset in males at 8 years old vs.14 years in females), through puberty. Studies reveal females have an increased risk relative to males during adulthood. Where sudden death occurred, males died on average at 13 years whereas females at 20 years. Males had a 32 % incidence of sudden death as a first episode in comparison to 9% in women. Studies showed that among referred subjects to the LQTS Registry, “only a few males had a first cardiac event after 15 years of age whereas about half of the females had a first cardiac event after 15 years of age.” (1)

The paper by Eric J. Rashba, et.al., “Influence of Pregnancy on Risk for Cardiac Events in Patients with Hereditary Long –QT Syndrome”, reviews the findings of a retrospective study of 422 LQTS women, (111 probands or first identified family member with LQTS and 311 first degree female relatives) who have experienced at least one or more pregnancies. Symptom information on these subjects was reviewed for the 40-week pregnancy period plus the 40 weeks prior to pregnancy and the 40 weeks post pregnancy or post-partum period. Analysis showed of the 111 probands, 26 (23.4%) had cardiac events during the post-partum period, with 10 (9%) during pregnancy and 4 (2.8%) in the pre-pregnancy period. (Only 2 patients had cardiac events during delivery with the remaining 24 experiencing events during the 40 weeks post-partum.) (2)

Thus, this study implicates that probands were at a significant risk for cardiac events during the post-partum period; additionally nearly 10% having a first cardiac event during this time frame. However, Beta-blocker treatment was also noted to associate with meaningful reduction of risk for cardiac events among probands. Also mentioned was that Beta-blockers studied extensively in hypertensive patients during pregnancy noted that maternal treatment with specifically “propranolol was infrequently associated with neonatal bradycardia, respiratory depression, hypoglycemia and intrauterine growth retardation in several small, uncontrolled studies....fetal malformations post maternal treatments with (propranolol, atenolol and metoprolol) were not observed.” (2; for further reading refer to referenced articles in this manuscript on Beta-blocker treatment with pregnancy.)

In over 10 years of retrospective communications with LQTS patients and family members post cardiac events during the post-partum period, (most have been cardiac arrests or sudden deaths), several commonalities have been noted:

1. All patients had their events in association with severe anxiety, illness, sleep deprivation, baby blues or depression. (Sleep deprivation being the most common or in combination with one or more previously named precipitators.)
2. Loud and unexpected noise---alarm clock, or sudden arousal from sleep.
3. Inattention to proper diet, poor eating habits or extreme weight loss or exercise.
4. Over the counter medications which were later discovered to prolong the QT-interval.

Suggestions for the 40-week post-partum period and beyond:

1. Maintain strict compliance with prescribed Beta-blocker medications. (3)
 2. Avoid all drugs or medications implicated to cause further prolongation of the QT-interval.
 3. Adhere to a healthy diet, and particularly increase the intake of potassium rich foods.
 4. Get plenty of rest.
 5. Remove all alarm clocks or telephones from resting areas. (3)
 6. Reduce, as much as possible, stress and anxiety.
 7. Seek help to relieve problems of depression, but be cautious in taking prescribed medications for depression.
 8. Consider the addition of omega-3 oils to diet. (4-6)
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1. Locati EH, et. al., "Age and Sex-Related Differences in Clinical Manifestations in Patients with Congenital Long-QT Syndrome", *Circulation* 1998; 97:2237-2244.
 2. Rashba EJ, et. al., "Influence of Pregnancy in the Risk for Cardiac Events in Patients with Hereditary Long-QT Syndrome", *Circulation* 1998; 97:451-456.
 3. Schwartz JP, et. al., "Genotype-Phenotype Correlation in the Long-QT Syndrome", *Circulation* 2001; 103:89-95.
 4. Kang JX, Leaf A, "Antiarrhythmic Effects of Polyunsaturated Fatty Acids", *Circulation* 1996; 94:1774-1780.
 5. Nair SSD, et. al., "Prevention of Cardiac Arrhythmia by Dietary (n-3) Polyunsaturated Fatty Acids and their Mechanism of Action", *Journal of Nutrition* 1997; 127:383-393.
 6. Leaf A, Kang JX, "Dietary n-3 Fatty Acids in the Prevention of Lethal Cardiac Arrhythmias", *Nutrition* 1997; 8:4-6.

Submitted by: Katherine Timothy
Clinical Coordinator of Cardiac Arrhythmia Genetic Research
Mark T. Keating Genetic Laboratory
University of Utah and Harvard University