

# Fears and Anxiety in Children with Long-QT Syndrome Compared to Children with Asthma

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**Objective:** To compare children with asthma to children with long-QT syndrome (LQTS) in terms of anxiety and medical fears.

**Method:** Forty children (25 males/15 females) with asthma and their mothers participated, along with seven children with LQTS (four males/three females) and their mothers.

**Results:** Children with asthma had significantly more medical fears, fear of danger/death, and fear of minor injury and small animals compared to children with LQTS. Children with LQTS tended to have more fear of failure and criticism, and tended to keep their feelings to themselves and minimize their real feelings of anxiety. Children with LQTS had significantly more internalizing problems, and their mothers had significantly higher anxiety.

**Conclusion:** Fear and uncertainty can be overwhelming in LQTS. Children with LQTS do not seem to be able to share their feelings openly. Examining the psychosocial adjustment of affected children may assist professionals to help families to cope more effectively.

**Key words:** psychiatry ■ QT intervals ■ asthma

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## INTRODUCTION

Long-QT syndrome (LQTS) is a constellation of repolarization disorders characterized by prolongation of the corrected QT interval on a standard electrocardiogram (ECG), T-wave abnormalities, relative bradycardia, conduction system disease and ventricular tachyarrhythmias.<sup>1</sup>

LQTS may cause sudden death in affected families, particularly in children and young adults who have not been diagnosed.<sup>1,2</sup> Parents may be advised by caregivers

to restrict their child's daily activities in order to avoid mental stress, strenuous sports and loud noises.<sup>3-5</sup> Adding to the stress of avoiding triggers is the fact that beta-blockers used for the treatment can cause side effects, such as mood swings, depression and fatigue, that may in turn result in noncompliance particularly for adolescent patients.<sup>1</sup>

There are surprisingly few studies examining the psychosocial aspects of LQTS syndrome. Preliminary studies of families screened for inherited cardiac arrhythmias have reported high levels of distress in parents of children at risk for LQTS.<sup>6</sup> Subsequent studies have confirmed the presence of clinically high levels of distress in parents whose children underwent predictive ECG testing and DNA testing for LQTS.<sup>7</sup>

A recent study examined parental perceptions about LQTS.<sup>1</sup> Parents wrote answers to seven open-ended questions about life with LQTS, including fear of death, quality of life, the impact of education on decisions about care and the impact on children. The 31 parents who participated had genotyped LQTS themselves, and only five parents had children <18 years of age. Two parents reported sudden cardiac death in their child at 18 years of age due to undiagnosed LQTS. Parents acknowledged the limited knowledge about LQTS in the general population and also among healthcare professionals, and said how challenging it was to have no one to talk to who would understand what living with LQTS is like. Of the 19 parents who expressed fears of death, all were worried about their children dying. Parents helped alleviate their fears by giving their children cell phones and frequently checking messages, using baby monitors in their child's room at night and taking defibrillators to every event that their children participated in.<sup>1</sup>

Parents and children with congenital heart disease are frequently exposed to extensive discussions regarding risks associated with progression of the disease process, risks of diagnostic interventions, and intraoperative and postoperative complications. For patients and families with LQTS, these discussions would also include risk of sudden death and measures to prevent its occurrence. Given the fact that the natural history of

LQTS is so extreme (ranging from no symptoms to sudden death), patients with LQTS and their families would be expected to experience significant anxiety and fears.

In contrast to LQTS, asthma is one of the most common chronic illnesses in children, and in Canada it is estimated that 13% of children and adolescents have asthma.<sup>8</sup> Asthmatic patients and their parents are often fearful of death, and asthmatic patients also exhibit an increased vulnerability toward the development of anxiety disorders.<sup>9</sup> Some children with asthma develop such severe anxiety with an attack that they panic, which often exacerbates the condition and interferes with treatment. In this current pilot study, this hypothesis was studied in detail by comparing children with asthma to children with LQTS in terms of anxiety and medical fears. We anticipated that children with LQTS would have significantly more anxiety and medical fears than children with asthma.

## METHODS

### Subjects

This study was approved by the research committee of the Alberta Children's Hospital and the Conjoint Medical Research Ethics Board, The University of Cal-

gary. Forty children (25 males, 15 females) from the asthma clinic participated in a previous study with the same questionnaire measures,<sup>10</sup> so their grouped data were used as a chronic illness comparison group for children with LQTS. None of the subjects in this study or the previous study of children with asthma had been identified as having any psychosocial needs.

All families followed at Alberta Children's Hospital cardiology clinic were contacted for enrollment and consent. There were five families who consented; seven children with LQTS and their mothers participated. There were four males and three females (average age 13.7 years, SD=2.1; range from 10–16 years). There were two sibling pairs participating (both were brother and sister sibling pairs).

In children with asthma, diagnosis was made by standard clinical criteria supplemented by spirometry before and after bronchodilator. A change of 15% in forced expiratory volume (FEV) was considered positive.<sup>11</sup> All children were on prophylactic high-potency inhaled steroids (budesonide, beclomethasone 250 mg/puff or fluticasone). These children at presentation required step-3 or step-4 therapy,<sup>11</sup> but with education, long-term follow-up and appropriate management strategies, symptomatic episodes became less common. Severity was

**Table 1. Sample characteristics**

| Variable                               | LQTS Group (N=7) |       | Asthma Overall Group (N=40) |       | Severe Asthma Subgroup (N=10) |       |
|--|------------------|-------|-----------------------------|-------|-------------------------------|-------|
|  | Mean             | SD    | Mean                        | SD    | Mean                          | SD    |
| Child's age (years)                    | 13.71            | 2.14  | 11.10                       | 2.88  | 12.13                         | 2.76  |
| Mom's age (years)                      | 42.71            | 6.07  | 39.48                       | 5.46  | 40.60                         | 2.97  |
| Fear of failure & criticism            | 35.43            | 6.43  | 29.40                       | 7.68  | 29.60                         | 7.79  |
| Fear of the unknown                    | 25.43            | 6.55  | 27.15                       | 6.50  | 26.70                         | 5.42  |
| Fear of minor injury and small animals | 24.00            | 6.32  | 32.08                       | 6.61  | 30.80                         | 5.29  |
| Fear of danger and death               | 21.00            | 5.48  | 25.95                       | 5.61  | 24.20                         | 5.75  |
| Medical fears                          | 5.71             | 1.98  | 9.53                        | 2.70  | 9.50                          | 3.31  |
| RCMAS total anxiety T score            | 49.14            | 10.12 | 50.58                       | 10.31 | 51.50                         | 10.99 |
| Physiological anxiety T score          | 10.57            | 3.82  | 10.90                       | 3.04  | 11.60                         | 3.72  |
| Worry T score                          | 8.71             | 3.86  | 8.98                        | 3.16  | 8.70                          | 4.00  |
| Social concerns T score                | 9.14             | 1.95  | 9.28                        | 2.78  | 9.60                          | 2.41  |
| Lie T score                            | 10.57            | 3.41  | 8.38                        | 2.95  | 9.60                          | 3.06  |
| STAI state anxiety standard score      | 47.57            | 8.04  | 33.43                       | 9.73  | 32.80                         | 14.07 |
| STAI trait anxiety standard score      | 49.29            | 8.44  | 32.59                       | 7.16  | 28.30                         | 5.48  |
| CBCL total T score                     | 56.43            | 7.74  | 53.50                       | 9.88  | 49.60                         | 6.72  |
| CBCL internalizing T score             | 63.29            | 10.67 | 55.23                       | 10.74 | 53.10                         | 8.28  |
| CBCL externalizing T score             | 50.86            | 5.37  | 50.60                       | 9.01  | 46.00                         | 4.55  |

determined by frequency of breakthrough attacks. Mild was <1 attack in 12 months, without interval symptoms; moderate was >1 attack in 12 months, without interval symptoms; and severe was persistent nighttime symptoms, exercise limitation and pulmonary function abnormalities between attacks.

In children with LQTS, diagnosis was established using ECG, Holter, treadmill and family history. In those patients who chose to undergo molecular testing, four patients were identified with LQTS type 2.

## Measures

Two child and two parent questionnaires were used. The child questionnaires were:

1. The Fear Survey Schedule for Children—Revised (FSSC-R). This instrument contains 80 items rated on a three-part scale for level of fearfulness. The instrument has five factored subscales. Fear of danger and death, and medical fears, are included in the factored subscales.
2. The Revised Children's Manifest Anxiety Scale (R-CMAS)<sup>12</sup>—a self-report measure consisting of 37 items to which the subject responds yes/no was used.

The parent questionnaires were:

1. The Achenbach Child Behavior Checklist (CBCL).<sup>13</sup> This factor is analysed into eight domains. In this study, the total T score, the internalizing T score and externalizing T score were considered only. T scores are commonly used standard scores and reflect how each subject's scores deviate from those for children of the same age range and sex from the normative sample. A T score of >67 is usually considered to be very much above average.
2. The State-Trait Anxiety Inventory<sup>14</sup> consists of 20 statements that evaluate how respondents feel "right now, at this moment." For this study, they were asked to respond specifically to "how I feel when I think of my child's health." The Trait-Anxiety Scale consists of 20 statements that assess how the respondent normally feels.

## RESULTS

### Comparisons with Children with Asthma

Results showed no significant group difference in age of mothers, but children in the LQTS group were significantly older than children in the asthma group [ $t(45)=2.48, p=0.027$ ] (Table 1). Children in the asthma group had significantly higher scores on the FSSC-R for medical fears [ $t(45)=-3.56, p=0.001$ ], fear of danger and death [ $t(45)=-2.36, p=0.036$ ], and fear of minor

injury and small animals [ $t(45)=-3.00, p=0.004$ ] compared to children in the LQTS group. Children in the LQTS group tended to have higher scores for fear of failure and criticism compared to children in the asthma group [ $t(45)=1.96, p=0.057$ ].

No significant differences emerged for anxiety in children overall on the R-CMAS or for physiological anxiety, worry or anxiety about social concerns. There was also a trend for children with LQTS to score higher on the lie subscale of the R-CMAS compared to children with asthma [ $t(45)=1.78, p=0.082$ ]. A high score on the lie subscale has been found to be equally useful in predicting anxiety problems as a high overall anxiety score on the R-CMAS (Reynolds & Richmond, 1985). The typical cutoff for the lie subscale is a standard score >13. There was a significant association between scoring >13 on the lie subscale and being in the LQTS group [ $\chi^2(1, N=47) = 6.78, p=0.009$ ]. In total, 28.6% of the children in the LQTS group scored above the cutoff on the lie subscale compared to only 2.5% in the asthma group.

There was a significant association between group and having a T score >67 for CBCL internalizing problems: 42.9% of children with LQTS had significantly elevated T scores for internalizing problems, compared to 10.0% of children with asthma [ $\chi^2(1, N=47)=5.07, p=0.024$ ].

As for mothers, results showed that mothers of children with LQTS had significantly higher State-Trait Anxiety scores [ $t(45)=3.63, p=0.001$ ] but no differences in Trait Anxiety scores compared to mothers of children with asthma. There were significant correlations between higher mother's state anxiety and higher scores for the child on the CBCL for internalizing problems.

Thus, having high scores on the lie subscale of the R-CMAS, on internalizing problems on the CBCL and on State-Trait Anxiety scores seemed to be more common in the LQTS group. The next analysis used the R-CMAS lie subscale, the internalizing problems from the CBCL and the State-Trait Anxiety in parallel screening, which involved identifying subjects with scores above the cutoff for  $\geq 2$  of these three tests. There was a significant association between being in the LQTS group and scoring above the cutoff on  $\geq 2$  of these tests [ $\chi^2(1, N=47) = 11.58, p=0.001$ ]. There were 57.1% of LQTS subjects scoring above the cutoffs on  $\geq 2$  of these tests compared to only 7.5% in the asthma group.

### Comparisons with Children with Severe Asthma

The final set of analyses involved comparing the children with LQTS to the subgroup of 10 children identified as having severe asthma. Children with severe asthma had significantly higher fears of minor injury or small animals [ $t(15)=-2.41, p=0.029$ ] and medical fears [ $t(15)=-2.69, p=0.017$ ] than children with LQTS. Mothers of children with LQTS had significantly higher state anxiety [ $t(15)=2.49, p=0.025$ ] and trait anxiety [ $t(15)=2.49, p=0.025$ ]

(15)=6.25,  $p<0.001$ ) than mothers of children with severe asthma. Children with LQTS tended to score above the cutoff for the lie subscale of the R-CMAS more often than children with severe asthma [ $\chi^2$  (1, N=17) = 3.24,  $p=0.072$ ]. Children with LQTS also had significantly higher T scores for internalizing problems [ $t$  (15)=2.22,  $p=0.042$ ] and tended to have higher T scores for externalizing problems [ $t$  (15)=2.02,  $p=0.062$ ] and for overall behavior problems [ $t$  (15)=1.94,  $p=0.072$ ] compared to children with severe asthma.

The final analysis used the R-CMAS lie subscale, the internalizing problems from the CBCL and the State-Trait Anxiety in parallel screening, as mentioned above. Once again, there was a significant association between being in the LQTS group and scoring above the cutoff on two or more of these three tests [ $\chi^2$  (1, N=17) = 4.41,  $p=0.036$ ]. There were 57.1% of LQTS subjects scoring above the cutoffs on  $\geq 2$  of these tests compared to only 10.0% in the severe asthma group.

## DISCUSSION

The goal of this study was to compare children with asthma to children with LQTS in terms of their anxiety and medical fears. We anticipated that the results from this study will be similar to those from our previous studies<sup>10</sup> where we found that physiological anxiety, medical fears and maternal anxiety are important issues requiring attention in asthma and cardiac disease, even in the absence of obvious psychosocial problems.

The present study showed that children in the asthma group had significantly higher scores on medical fears, fear of danger and death as well as fear of minor injury. It would be reasonable to assume that the reason is because children with asthma often experience physical sensation of not being able to breathe (which is no doubt a frightening sensation), whereas children with LQTS often do not experience any physical sensations and are only aware of their problem after the first episode of syncope.

The results also showed a trend for children with LQTS to score higher on the lie subscale of the R-CMAS. Research in this area has shown that lie scale is higher in children who express a high need for social desirability.<sup>15</sup> We have also learned that these children exhibit high scores on fear of failure and criticism. Therefore, it seems likely that the lie score is a means of compensating for their actual fears of failure, criticism and the need for social acceptance.

Despite our small sample of children with LQTS, the results also indicate that the children with LQTS score high on both internalizing and externalizing behaviors. It is possible that these children are not able to discuss feelings of anxiety and in turn cannot cope with their anxiety in a more open manner by sharing these feelings. This perhaps results in withdrawn or acting-out behaviors. Therefore, consideration of these findings may be

important when treating children with LQTS.

The potential of using parallel screening of high scores on the lie subscale of the R-CMAS, on internalizing problems on the CBCL and for maternal anxiety on the State-Trait Anxiety scale is also important. Scoring above the cutoff on  $\geq 2$  of these tests occurred in 57% of cases in the LQTS group compared to  $\leq 10\%$  for children with asthma. All three of these tests would be easily administered and scored in a clinic setting to help identify children and families at risk of problems adjusting to LQTS.

Parental anxiety about their child's illness has an impact on many factors, including clinical judgement, understanding of the condition and compliance with treatment.<sup>16</sup> Fears and uncertainty in LQTS can be overwhelming for the entire family when the diagnosis is first made.<sup>1</sup> Our finding of significantly higher state anxiety in mothers of children with LQTS confirms previous studies of families screened for inherited cardiac arrhythmias where high levels of distress in parents of children at risk for LQTS and/or undergoing predictive testing for LQTS have been reported.<sup>6,7</sup> Parental distress was particularly elevated in parents who were familiar with LQTS for a longer time, who had more experiences with LQTS in their family members and who received positive test results for their children.

Having a child with a chronic illness makes considerable emotional and practical demands on all family members, not just mothers.<sup>17</sup> Given the nature of this syndrome and the profound effect of the diagnosis on the individual and the family, future studies should consider adjustment and well-being in children with LQTS and their families. Much of the previous research into the adjustment of chronically ill children, their parents and their siblings has focused on illnesses such as cancer, cystic fibrosis and spina bifida.<sup>17</sup> Unfortunately, similar research on pediatric cardiology patients in this regard is lacking. Examining the psychosocial adjustment and well-being in families of children with LQTS may assist cardiologists, psychologists and clinical geneticists to identify families at risk for problems adjusting to LQTS, and to assist the families of children with LQTS to cope more effectively with this syndrome.

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