

BRIEF COMMUNICATION**INCIDENCE AND CHARACTERISTICS OF LYMPHEDEMA
IN TURNER'S SYNDROME****J. Welsh, M. Todd**

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ABSTRACT

A survey of patients with Turner's syndrome and their parents documents the incidence, features, and prognosis of lymphedema in surviving children.

Keywords: Turner's syndrome, congenital lymphedema, lymphatic dysplasia, children, survey

The reported incidence of lymphedema at birth in Turner's syndrome (TS) ranges from 30-51% (1-3). The swelling is thought to be temporary in approximately 80% of children but persists in some in the lower legs (4). A questionnaire was distributed to 200 participants of the National Turner's Syndrome Conference in Blackpool. The conference was attended by individuals who were affected by Turner's and their parents. A total of 74 questionnaires were returned. Lymphedema was present in 57% (n=42) and not present in 43% (n=32). Of those with lymphedema, 76% (n=32) had swelling

at birth, 19% (n=8) developed some time later than birth and 2 were unsure. The majority of respondents 55% (n=23) reported swelling in hands and feet (*Fig. 1*). Swelling had resolved totally in 19% (n=8), improved in 31% (n=13), worsened in 10% (n=4), and remained unchanged in 40% (n=17). Where swelling had completely resolved, this occurred by the age of 2 years in 87% (n=7).

Sixty-five percent reported receiving no treatment at all for the swelling, and respondents attributed this lack of treatment to doctors' failure to appreciate the impact of the swelling and a lack of knowledge that effective treatment is available. Half of those receiving treatment reported difficulty in accessing treatment. Psychological problems included frustration with the swelling, altered body image, being teased at school and feeling low in mood because of the swelling.

This survey gives an incidence of lymphedema in surviving children with TS at 57%. Contrary to information in textbooks,

there is some evidence from this small study that lymphedema persists in the majority (81%) of individuals with TS. A larger study is required to validate these findings and provide parents and children with more reliable information on the incidence of lymphedema and its prognosis.

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