The Familial Incidence of Carpal Tunnel Syndrome in Patients With Unilateral and Bilateral Disease

J. Winslow Alford, MD, Arnold-Peter C. Weiss, MD, and Edward Akelman, MD

Abstract

The controversy has been considerable regarding the incidence of familial carpal tunnel syndrome. Of particular interest is the relationship between bilateral disease and familial incidence. In this study, we compare the incidence of familial carpal tunnel syndrome in patients with the syndrome in one, both, or neither hands.

We report a significantly greater incidence of familial carpal tunnel syndrome in those patients with bilateral disease than in a similar population of patients with either unilateral disease or no carpal tunnel syndrome. These results may imply inheritability of variations in the size of the tunnel or its contents, which would manifest themselves bilaterally and may cause a predisposition for developing the syndrome. Being aware of the propensity for bilateral disease may improve our ability to prevent and treat the disease.

Carpal tunnel syndrome refers to the diagnosis of median nerve entrapment at the wrist as the median nerve travels through the carpal tunnel. The carpal tunnel is a narrow channel on the volar aspect of the carpus through which run the median nerve and all of the extrinsic flexor tendons. The tunnel is bounded dorsally by the carpal bones, ulnarly by the hook of the hamate bone, radially by the trapezium, and anteriorly by the transverse carpal ligament.1 Median nerve compression may occur with any increase in the size of the contents in the tunnel, decrease in the size of the tunnel itself, or increase in the pressure within the tunnel. Symptoms associated with median nerve compression have been recognized for more than a century, but carpal tunnel syndrome was not defined as a clinical entity until Phalen and colleagues’ work between 1950 and 1970.2,3 Despite widespread knowledge of this syndrome and its treatment, a high incidence of carpal tunnel syndrome persists.4 In addition, numerous reports indicate carpal tunnel syndrome trends within families.5-11 These trends are associated with a host of defects, ranging from systemic biochemical aberrations to inheritable structural anomalies of the carpal tunnel itself.

Biochemical changes associated with familial carpal tunnel syndrome include familial amyloidosis polyneuropathy as a result of transthyretin variants from various point mutations.5,6 Systemic disorders have included inheritable myopathies7 and familial hypercholesterolemia.8 Structural irregularities have also been reported. An example of an often reported structural anomaly associated with familial carpal tunnel syndrome is a thickened transverse carpal ligament,9 including 1 report of a surgically identified median nerve aplasia distal to but not proximal to a thickened transverse carpal ligament in a 7-year-old who had 3 immediate family members with identical abnormalities.10 Other structural changes associated with familial carpal tunnel syndrome include a congenitally small carpal tunnel, distal prolongation of the superficial flexor muscle bellies, anomalous muscles, and anomalous paths of the medial artery and median nerve branches.11

These reports of inheritable irregularities and the coexistence of carpal tunnel syndrome have stimulated considerable work to examine the extent to which inheritable traits play a role in the development of carpal tunnel syndrome. To date, little agreement exists as to the inheritability of carpal tunnel syndrome. As early as 1959 Tanzer12 described carpal tunnel syndrome as a “familial trait” in 4 of 22 (18%) surgical patients. In 1966 Phalen13 reported a prevalence of family occurrence, adding that many patients volunteer that all their relatives have the same constellation of symptoms, and concluded that “there is probably some familial predisposition to carpal tunnel syndrome.” In 1975 Danta9 suggested that a
family occurrence was more common than previously thought by reporting that children with carpal tunnel syndrome often had a family member with the same symptoms. An opposing opinion from Stevens and colleagues revealed no dramatic trends within families in their analysis of conditions associated with carpal tunnel syndrome. Most recently, however, Radecki demonstrated an increased incidence of family occurrence in patients with carpal tunnel syndrome. Radecki’s findings, however, may have been confounded because persons who are diagnosed with carpal tunnel syndrome become more aware of family members with the syndrome and are more likely to report that they have carpal tunnel syndrome in their families. In addition, the paper did not differentiate between bilateral and unilateral disease.

If inheritable carpal tunnel syndrome is caused by biochemical, developmental, or anatomical changes of the carpal tunnel, then these changes are likely to occur bilaterally. We thought it would be useful then to conduct an analysis of familial carpal tunnel syndrome with respect to the incidence of bilaterality.

The purpose of this study was to compare the incidence of familial carpal tunnel syndrome in unilateral and bilateral disease. We assessed the frequency of familial occurrence of carpal tunnel syndrome in patients with and without carpal tunnel syndrome. By comparing the incidence of familial carpal tunnel syndrome in unilateral and bilateral carpal tunnel syndrome patients, we hoped to control for the “education factor” that has caused difficulty in previous studies.

Materials and Methods
To assess the familial occurrence of carpal tunnel syndrome in patients with carpal tunnel syndrome compared with similar patients without carpal tunnel syndrome, we conducted a randomized, case-controlled retrospective study on 161 patients in our orthopedic hand surgery offices. All patients with regularly scheduled appointments in our upper extremity orthopedic offices completed a survey that, in addition to demographic information, identified patients with carpal tunnel syndrome. No limitation was placed on their motivation for making the appointment. Additional information included questions about the patient’s age, gender, occupation, dominant hand, risk factors for carpal tunnel syndrome, hand symptoms, and affected hand(s). In addition to occupation, specific risk factors considered in the survey were diabetes mellitus, rheumatoid arthritis, thyroid dysfunction, pregnancy, and any hobbies that amounted to greater than 3 h/wk and involved a significant amount of repetitive wrist motion. Carpal tunnel syndrome patients were asked to indicate which treatments they had received for their carpal tunnel syndrome (ie, bracing, steroid injections, or surgical release of the carpal tunnel). All participants were then asked whether they knew of one or more family members with carpal tunnel syndrome, and identical information about risk factors for these family members was obtained. All information was self-reported.

Any patient who reported the aforementioned medical risk factors for carpal tunnel syndrome was eliminated from the study. The remaining 144 patients were entered into the study and analyzed using Instat (Graphpad Software, Inc, 1998). A chi-square test for trend was conducted because the number of hands affected (0, 1, or 2) is a natural order of increasing involvement.

Results
The population consisted of 161 patients with regularly scheduled appointments in our hand surgery offices. Seventeen patients had previously diagnosed medical conditions, resulting in elimination from the study. They are indicated in Table I. The remaining 144 patients had an average age of 39.4 (range, 17 to 398 AUGUST 2004

![Figure 1](https://example.com/figure1.png)

**Figure 1.** Number of patients with unilateral and bilateral carpal tunnel syndrome (CTS) and the number of family members with carpal tunnel syndrome.

![Figure 2](https://example.com/figure2.png)

**Figure 2.** Percentage of patients with unilateral and bilateral disease with familial carpal tunnel syndrome (CTS).
80, SD = 12) with 42 male and 102 female patients. Of the 144 patients, 111 were right-hand dominant, 26 were left-hand dominant, and 7 considered themselves “ambidextrous.” Although a greater number of female patients had the syndrome, women were no more likely to have bilateral disease (Table II).

The data were then cross-tabulated to analyze the relationship between the number of hands affected and the existence of one or more family members with carpal tunnel syndrome. The correlation table for these variables is illustrated in Table III.

A chi-square test for independence showed a chi-square of 7.61 with 2° of freedom, and $P = 0.022$. For this analysis the chi-square for trend is 7.23 with 1° of freedom, and $P = 0.007$. Figure 1 displays these results. Forty-five percent of the patients (30/67) with bilateral disease had family members with carpal tunnel syndrome, whereas only 27% (7/26) of the unilateral carpal tunnel syndrome patients had family members with carpal tunnel syndrome. Among the 51 patients without carpel tunnel syndrome in either hand, only 11 (22%) reported family members with the syndrome.

**Discussion**

Self-reporting of the data placed unavoidable limitations on the accuracy of our data because we relied on patients’ awareness of their family members’ conditions. By comparing unilateral and bilateral carpal tunnel syndrome patients, we attempted to control for over-reporting by carpal tunnel syndrome patients who may have a greater awareness of family members’ carpal tunnel syndrome status. However, we were not able to assess the degree to which non–carpal tunnel syndrome patients were underreporting family members with carpal tunnel syndrome of whom they were unaware.

Despite these limitations, our data suggest a relationship between bilateral carpal tunnel syndrome and familial carpal tunnel syndrome, including an intermediate relationship in those patients with unilateral carpal tunnel syndrome (Figure 2). Most familial carpal tunnel syndrome occurs in patients with bilateral disease.

Of particular interest is to place our data in perspective with what is currently understood about familial carpal tunnel syndrome. In 1994 Radecki reviewed 421 patients and reported a 39.3% rate of familial carpal tunnel syndrome in patients with prior carpal tunnel release versus 13.3% in patients without electromyogram-diagnosed carpal tunnel syndrome. These data agree with our findings, but our analysis adds another level of understanding. Radecki’s rate of approximately 40% lies between our rate of 27% for unilateral disease and 45% for bilateral disease. Because that study’s data do not differentiate between bilateral and unilateral carpal tunnel syndrome patients, we will not know whether perhaps the majority of the familial carpal tunnel syndrome patients were in fact bilateral carpal tunnel syndrome patients.

The high incidence of familial carpal tunnel syndrome in patients with bilateral disease may suggest either a systemic biochemical irregularity or an inheritable structural variation in the size of the tunnel or the
volume of its contents. Of interest for further investigation would be the age of onset of carpal tunnel syndrome in the patients and their family members.

What is not clear from our data is whether familial carpal tunnel syndrome is a result of congenital, developmental, or environmental factors. A prospective study following younger members of the carpal tunnel syndrome families could assess the role of developmental changes in the acquisition of carpal tunnel syndrome. Alternatively, diagnostic imaging of the precise carpal tunnel architecture in patients and their family members could clarify the role of specific anatomical irregularities in the development of carpal tunnel syndrome.

**Conclusion**

Our results demonstrate a relationship between bilateral carpal tunnel syndrome disease and the existence of familial carpal tunnel syndrome. This relationship is not as pronounced in unilateral disease, which eliminates the possibility that bilateral carpal tunnel syndrome patients are overreporting family members because of a greater awareness of carpal tunnel syndrome in their families.

Bilateral carpal tunnel syndrome may be used as an indication to monitor family members carefully for the development of carpal tunnel syndrome. Our results raise the possibility of inheritable anatomical variation that manifests bilaterally and causes a predisposition for developing the syndrome. A heightened awareness of carpal tunnel syndrome in the other members of families with a bilateral carpal tunnel syndrome patient may allow for more effective preventive measures through ergonomic modifications at work or through earlier and more aggressive treatment options.

**References**