

FAMILIAL BILATERAL CARPAL TUNNEL SYNDROME IN A NIGERIAN FAMILY: CASE REPORT

Johnson D. Ogunlusi, FMCS (Ortho);¹ Lawrence M. Oginni, FMCS, FWACS²

CASE PRESENTATION

In a family in the southwestern part of Nigeria, eight out of nine siblings had classical features of bilateral carpal tunnel syndrome (CTS). Their mother, first cousin, and an aunt also had bilateral involvement of their hands. They all presented similarly with nocturnal wrist pain and paresthesias in the media nerve territory. Their occupations all varied, but included physician, teacher, engineer, zoologist, businessman and mechanic.

In total, three male and five females were affected. All were right handed and all were above 40 years of age at the time of presentation. Medically, the oldest of the patients had hypertension. Otherwise, there was no family or patient history of diabetes mellitus, myxedema, obesity, gout or rheumatoid arthritis. They were all generally fit and muscularly built. There was thenar muscle wasting in one patient (the medical doctor) who presented late. Five of the siblings underwent release of the bilateral carpal tunnels at one sitting. Four surgeries were performed within five months of onset of symptoms, and one at 14 months of onset of symptoms. The releases were all done by the same orthopaedic surgeon under tourniquet and involving the vertical division of the flexor retinaculum and transverse carpal ligament under direct visualization, through a 5-cm skin incision. The incision extended from the junction of proximal third and distal two-thirds of the thenar crease, along the ulnar border of the thenar crease, almost to the transverse skin crease of the wrist. Bandaging with collar and cuff was applied for 14 days. All the wounds

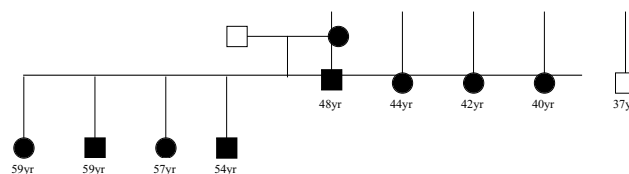


Figure 1. Family tree showing siblings and mother affected with CTS, and their ages at presentation.

healed primarily. Splinting was not done. Postoperative physiotherapy was performed after the incisions healed for two weeks, and for six months for the patient with thenar atrophy who presented late. There was complete recovery of the neuromuscular function of the hands in the four patients that presented early for surgery and there was residual atrophy in the patient that presented late with thenar atrophy. The history and the management outcomes of the patients are shown in Table 1.

DISCUSSION

The literature reveals that familial bilateral carpal tunnel syndrome may be a genetically distinct disorder.^{1,2} In some reports, onset is in childhood.³

Familial bilateral carpal tunnel syndrome has been reported in siblings with mucopolysaccharidosis III (pseudo-Hurler polydystrophy).⁴ Hereditary neuropathy with liability to pressure palsies (HNPP) is also a known cause of bilateral carpal tunnel syndrome in childhood, where the CTS may be the first manifestation of this systemic nerve pathology.⁵ Swoboda et al. reported a case of a seven-month-old infant with familial CTS presenting with mutilated hands due to recurrent chewing of his digits in a median nerve distribution.⁶ Gray et al. identified bilateral carpal tunnel syndrome in 19 out of 43 living persons of a nonconsanguineous family with no single common etiologic feature identified.¹ They suggested that an inheritable disorder transmitted by an autosomal dominant gene with high-degree penetrance might be responsible. Leifer et al. in Massachusetts reported two families with multiple members who had bilateral carpal tunnel syndrome, with a pattern consistent with autosomal dominance inheritance.⁷ Stoll et al. in

¹Department of Orthopaedics and Traumatology
Obafemi Awolowo University Teaching Hospitals Complex
Ile-Ife, Nigeria

²Department of Orthopaedics Surgery and Traumatology
Ile-Ife, Nigeria

Correspondence:

Dr. J. Ogunlusi
Department of Orthopaedics Surgery and Traumatology
College of Health Sciences
Obafemi Awolowo University
Ile-Ife, Osun State Nigeria
Telephone: 23408055336926
Telefax: 23436230141
Email: gbemidare@yahoo.com

TABLE 1
The History and Management Outcome Bilateral Carpal Tunnel Syndrome in a Family

S/N in the family	Age at presentation	Sex	Occupation	Associated disease	T M W at presentation	Bilateral Ct Release	Duration between onset & surgery	Outcome of Rx
1	59	F	Medical Practitioner	Hypertension	Present	+	14 months	Residual thenar muscles atrophy
2	59	M	Trading	Nil	Absent	+	4 months	Good Recovery
3	57	F	Teaching	Nil	Absent	-	-	-
4	54	M	Technician	Nil	Absent	+	2 months	Good Recovery
5	48	M	Agric Engineer	Nil	Absent	-	-	-
6	44	F	Teaching	Nil	Absent	+	4 months	Good Recovery
7	42	F	Teaching	Nil	Absent	+	3 months	Good Recovery
8	40	F	Zoologist	Nil	Absent	-	-	-

KEY

+ = Carpal Tunnel Release
 - = No Carpal Tunnel Release yet
 T M W= Thenar Muscles Wasting

Strasbourg, France reported a family with multiple early symptom onset with CTS between ages nine and 52 years of age, with an autosomal dominant inheritance pattern.⁸ Braddom reported familial carpal tunnel syndrome, not bilateral, in seven members of three generations of a black African-American family in United States.⁹ The ages of those affected ranged from 29 to 67 years.

Based on our search of the English-language literature, no case series of familial bilateral CTS has been reported in Africans in the literature. In our patients, traditional open carpal tunnel releases were performed. Bilateral releases were done at one surgical sitting to minimize the overall convalescence time, and surgical expenses. Bilateral releases have been reported to have acceptable outcomes in the literature.^{10,11,12} It is also noteworthy that our patients presented with bilateral carpal tunnel symptoms at ages above forty. No genetic testing was performed for our patients. In general, our case and the literature reveal that familial bilateral carpal tunnel syndrome with autosomal dominant penetrance could present in patients in adulthood, not necessarily in children. The family history and past medical history of these patients needs to be evaluated in detail. In our case, there were no other obvious underlying familial medical conditions. Further basic science and genetic studies in such patients with familial bilateral carpal tunnel syndrome may someday help better understand the role of HNPP or some other underlying disorder.

REFERENCES

1. **Gray RG, Poppo MJ, Gottlieb NL.** Primary familial bilateral carpal tunnel syndrome. *Ann Intern Med.* 91(1):37-40, Jul. 1979.
2. **Vallat JM, Dunoyer J.** Familial carpal tunnel syndrome [Article in French]: *Sem Hop.* 54(17-20):661-2, Jun. 1978.
3. **Danta G.** Familial carpal tunnel syndrome with onset in childhood: *J Neurol Neurosurg Psychiatry.* 38(4):350-5, April 1975.
4. **Starreveld E, Ashenurst EM.** Bilateral carpal tunnel syndrome in childhood. A report of two sisters with mucopolidosis III (pseudo-Hurler polydystrophy). *Neurology.* 25(3):234-8, March 1975.
5. **Cruz-Martinez A, Arpa J.** Pediatric bilateral carpal tunnel syndrome as first manifestation of hereditary neuropathy with liability to pressure palsies (HNPP). *Eur J Neurol.* 5(3):316-317, May 1998.
6. **Swoboda KJ, Engle EC, Scheindlin B, Anthony DC, Jones HR.** Mutilating hand syndrome in an infant with familial carpal tunnel syndrome. *Muscle Nerve.* 21(1):104-11, Jan. 1998.
7. **Leifer D, Cros D, Halperin JJ, Gallico GG 3rd, Pierce DS, Shahani BT.** Familial bilateral carpal tunnel syndrome: report of two families. *Arch Phys Med Rehabil.* 73(4):393-7, April 1992.
8. **Stoll C, Maitrot D.** Autosomal dominant carpal tunnel syndrome. *Clin Genet.* 54(4):345-8, Oct. 1998.
9. **Braddom RL.** Familial carpal tunnel syndrome in three generations of a black family. *Am J Phys Med.* 64(5): 227-34, Oct. 1985.

10. **Wang AA, Hutchinson DT, Vanderhooft JE.** Bilateral simultaneous open carpal tunnel release: a prospective study of postoperative activities of daily living and patient satisfaction. *J Hand Surg [Am]*. 28(5): 845-8, Sep. 2003.
11. **Weber RA, Boyer KM.** Consecutive versus simultaneous bilateral carpal tunnel release. *Ann Plast Surg*. 54(1): 15-9, Jan. 2005.
12. **Wilson JK, Sevier TL.** A review of treatment for carpal tunnel syndrome. *Disabil Rehabil*. 4; 25(3):113-9, Feb. 2003.