Carpal tunnel syndrome in children

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Carpal tunnel syndrome (CTS) is rarely seen in children. A literature search in 1989 revealed 52 published cases. The authors review 163 additional cases that were published since that date. The majority of these cases were related with a genetic condition. The most common aetiology was lysosomal storage disease: mucopolysaccharidoses (MPS) in 95 and mucolipidoses (ML) in 22. In CTS secondary to MPS, clinical signs typical of adult CTS are rarely seen, and difficulty with fine motor tasks is the most frequent finding. CTS in MPS does not seem to be prevented by bone marrow transplantation, the usual treatment for the condition. CTS is probably due to a combination of excessive lysosomal storage in the connective tissue of the flexor retinaculum and a distorted anatomy because of underlying bone dysplasia. Mucolipidoses come next in the aetiology, with essentially similar symptoms. The authors found in the literature 11 cases of primary familial CTS, a condition which presents as an inheritable disorder of connective tissue mediated by an autosomal dominant gene; the symptoms may be more typical in some cases, but are more similar to MPS in others. A case with self-mutilation has been reported. Hereditary neuropathy with liability to pressure palsies (HNPP) is a rare autosomal dominant condition characterised by episodes of decreased sensation or palsies after slight traction or pressure on peripheral nerves; it may also give symptoms of CTS. Schwartz-Jampel syndrome (SJS), another genetic disorder with autosomal recessive skeletal dysplasia, is characterised by varying degrees of myotonia and chondrodysplasia; it has also been noted associated with CTS in a child. Melorheostosis and Leri’s syndrome have also been noted in children with CTS, as well as Déjerine-Sottas syndrome and Weill-Marchesani syndrome. Among non-genetic causes of CTS in children, idiopathic cases with children onset have been reported, usually but not always related with thickening of the transverse carpal ligament. Intensive sports practice has been reported as an aetiologcal factor in several cases of childhood CTS. Nerve territory oriented macrodactily, a benign localised form of gigantism, is another unusual cause of CTS in children, as are fibrolipomas of the median nerve or intraneural perineuroma or haemangiom of the median nerve. Acute cases have been reported in children with haemophilia, secondary to local bleeding. Another local cause is a musculotendinous malformation of the palmaris longus, the flexor digitorum superficialis, the flexor carpi radialis brevis (a supernumerary muscle), the first lumbricalis or the palmaris brevis. Isolated cases of childhood CTS have also been reported in Klippel-Trenaunay syndrome, in Poland’s syndrome and in scleroderma. Finally, several cases have been noted following trauma, most often related with epiphysiolysis of the distal radius. Immediate reduction has cleared the problem in most cases, but exploration of the median nerve should be considered otherwise, and also in cases with delayed occurrence of symptoms. Overall 145 of the 163 reviewed cases have undergone open carpal tunnel release. Childhood CTS often has an unusual presentation, with modest complaints and children are often too...
young to communicate their problem. In CTS with specific aetiologies such as storage disease, the symptoms may be masked by the skeletal dysplasia and joint stiffness. Every child with even mild symptoms must be thoroughly examined and a family history must be taken. Children with storage disease may benefit from early clinical and electrophysiological screening before they develop obvious clinical signs.

Carpal tunnel syndrome (CTS) is frequent in adults, but the condition is rare during childhood. CTS was first reported in childhood by Martin and Masse, who recorded three cases in 1958 (26). All three children suffered from recurrent attacks of severe pain in the hand. No cause was identified in anyone of the patients. Two of them were helped by surgical decompression, whereas the other patient’s symptoms were relieved by peroral administration of corticosteroids.

The literature was thoroughly reviewed in 1989 by Poilvache et al (30). They reported 52 cases with varying aetiologies, with mucopolysaccharidosis and mucolipidosis being the most frequent cause of CTS in childhood (18 of 52 cases).

We reviewed 163 cases of paediatric CTS reported in the literature since 1989 (table I). The majority of the articles deal with case reports and small series. A search in Pubmed® was performed: since 1989, 35 articles on childhood CTS were retrieved. A review paper by Lamberti and Light was published recently (25). However this is far from complete in case finding.

The majority of cases of childhood CTS reported were related with a genetic condition. The most common aetiology, as reported in other articles, was lysosomal storage disease. Of the 163 reviewed cases, a total of 95 cases of CTS were due to mucopolysaccharidosis (MPS), and 22 cases to mucolipidosis (ML), which gives a total of more than 70% of the cases related with storage diseases (5-20). In the survey of Poilvache et al, 18 out of 52 (35%) patients with CTS had MPS or ML (30).

Mucopolysaccharidoses are inherited intracellular lysosomal storage disorders, caused by the deficiency of a specific lysosomal enzyme, in which the deposition of glycosaminoglycans leads to cell, tissue and organ dysfunction (table II) (18).

Hurler’s syndrome (type I H MPS) is the most common and most severe form in the group of MPS. It is inherited as an autosomal recessive deficiency of α-L-iduronidase. The function of this enzyme is to degrade the glycosaminoglycans (dermatan and heparan sulfate). When untreated, this results in coarse facial features, hepatosplenomegaly, severe mental retardation, and cardiovascular and pulmonary failure. Since the 1980’s treatment for Hurler syndrome has consisted of bone marrow transplantation (BMT).

According to Haddad et al, symptoms of CTS were rare in 42 MPS-patients, while signs such as decreased sweating, pulp atrophy, thanar muscles wasting and manual clumsiness were much more common. They found that numbness, tingling, nocturnal pain, Tinel’s sign and Phalen’s sign were often absent (18).

Van Heest et al (38) also found that in 17 MPS-patients with childhood CTS the most frequent complaint was notable difficulty with fine motor tasks. Wraith and Alani (40) included 14 patients with MPS in their study about CTS. No patient or parent reported symptoms suggestive of carpal tunnel syndrome, such as paresthesias or nocturnal pain. All patients with the exception of those under the age of two years at testing had clear evidence of wasting of the thanar muscles and of typical claw hand deformity seen in the MPS and related disorders. They concluded that carpal tunnel syndrome in MPS and related disorders is probably due to a combination of excessive lysosomal storage in the connective tissue of the flexor retinaculum as well as a distorted anatomy because of underlying bone dysplasia (40).

While the therapy of severe forms of MPS consists of bone marrow transplantation, the question could arise whether or not this would prevent CTS. In the study of Guffon et al (16), three out of eight patients with CTS due to MPS, had CT release before bone marrow transplantation, while the other five underwent surgery between one and nine years after bone marrow transplantation. Van Heest et al also wrote that all 12 MPS patients with CTS, who had not yet had carpal tunnel release, underwent surgery after bone marrow transplantation (38).
Lamberti and Light (25) noted that the median nerve in MPS does not seem to be profoundly involved in cases with CTS. The surrounding tenosynovium is swollen along with the tendons, and the transverse carpal ligament is consistently thickened. The articles we reviewed seem to be consistent with these findings. Haddad et al (17) described the flexor tendons in four MPS-patients with CTS and triggering in the wrist as a large white shiny mass of synovitis. In another survey Haddad et al (19) reported on 48 children with MPS or ML with CTS. The operative findings described are a typically very thickened flexor retinaculum over a large mass of shiny white tenosynovium, which engulfed the flexor tendons and restricted both single tendon and differential motion. In their cases there was a variable amount of nerve constriction and a thickened epineurium. Van Heest et al (38) found a thickened transverse carpal ligament with significant flexor tenosynovial deposits, resulting in an hourglass constriction of the median nerve in their survey of 17 MPS-patients with CTS. Biopsy of the tenosynovial tissue confirmed abundant proteoglycan deposition.

Next to mucopolysaccharidoses, mucolipidoses (ML) are a frequent cause of childhood CTS. These are similar disorders caused by an inherited deficiency of the enzyme necessary for complete post-translational processing of lysosomal hydrolases (table III). The clinical features produced by this enzyme deficiency mimic those of some of the mucopolysaccharidoses (40).
Type III mucolipidosis – also called “pseudo-Hurler” – is a frequent cause of CTS in children. The clinical presentation is usually heralded by upper limb stiffness between ages two and four years. A claw hand deformity develops by the age of six in association with CTS in many cases. There is usually a mild coarsening of facial features similar to that seen in the mucopolysaccharidoses. Intelligence is often reduced but can be normal (18).

The presentation of CTS in these children – sometimes the first presenting symptom of mucolipidosis - mimics the presentation in children with MPS who have CTS. The symptoms described are weakness of grip, paresthesias, wasting of thenar muscles, pain, static 2-point discrimination larger than 7 mm, decreased sweating and one case report describes self-mutilation in an ML-patient with CTS (41).

In the reviewed literature we found 11 cases of primary familial CTS. In familial CTS the age of onset is early and the sex distribution is nearly even. In families with CTS, onset often occurs at less than 40 years of age and sometimes in childhood. These findings, as well as probable father-to-son transmission earlier described, are consistent with an inheritable disorder of connective tissue mediated by an autosomal dominant gene (34).

Lamberti and Light (25) reported that symptoms of familial CTS in children are more typical with pain, numbness, and nocturnal paresthesias. In one of our patients symptoms were indeed rather typical, with nocturnal complaints of his fingers “going dead” and a sensation of “pins and needles” (11).

Michaud et al (27) however described a boy without typical complaints who lacked thumb opposition, interfering with the development of his fine motor skills and associated with a clumsy use of a pencil. He also demonstrated a preference for poking with his isolated fourth finger rather than the index finger. Physical examination of the patient revealed bilateral decrease in prominence of the thenar eminence, lack of opposition, and decrease in grip and pinch strength. Sensation was absent or severely impaired over the palmar surface of the three radial digits.

Swoboda et al (35) described a mutilating hand syndrome in a seven-month-old boy, the son of consanguineous parents. He presented with recurrent chewing of his digits in the median nerve distribution as the primary manifestation of CTS, in conjunction with features consistent with congenital insensitivity to pain. Bilateral carpal tunnel release led to complete cessation of hand mutilation and full recovery of fine motor function.

The only abnormality described so far in primary familial CTS has been the noninflammatory thickening of the transverse carpal ligament, which compresses the underlying nerve. The aetiology of this thickening is unclear (37).

Hereditary neuropathy with liability to pressure palsies (HNPP) is another entity also responsible for familial cases of CTS. It is an autosomal dominant disease characterised by episodes of decreased perception or palsies after slight traction or compression of peripheral nerves. Clinical features are usually noticed during the second, third or later decades of life. Some palsies however may start at an earlier age.

One case was described by Cruz Martinez and Arpa (10): a 5-year-old boy complaining of bilateral painful numbness over the digits innervated by both median nerves, of three months duration. Nerve conduction studies of the patient’s mother and sister showed abnormalities consistent with an inherited disorder of connective tissue mediated by an autosomal dominant gene (34).

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Schwartz-Jampel syndrome (SJS) is a genetic disorder that has been associated with CTS in childhood. SJS is an autosomal recessive skeletal dysplasia characterised by varying degrees of myotonia and chondrodysplasia. It features short stature, osteochondrodysplasia, myotonia, and a characteristic face with a “fixed” facial expression, blepharophimosis, pursed lips, and, sometimes, low-set ears and myopia. Skeletal abnormalities include kyphoscoliosis, platyspondyly with coronal clefts in the vertebrae, metaphyseal and epiphyseal dysplasias, and joint contractures. It is caused by a mutation in the perlecan gene, responsible for the synthesis of perlecan, a large heparan sulfate proteoglycan. The molecular mechanism underlying the chondrodystrophic myotonia phenotype of SJS is unknown (3). In the reviewed literature, we found one case with SJS and CTS: a 7-year-old girl without symptoms of CTS, but with nerve conduction studies showing bilateral abnormalities of the median nerves at the wrist (10). CTS seems to be secondary to the dwarfism, with narrow carpal structures and muscle hypertrophy, along with constant flexion of the wrist (10).

Melorrheostosis and Leri’s pleonosteosis are two rare disorders that have been associated with CTS. Melorrheostosis or osteosis eburnicans monomelica, inherited as a dominant trait, can cause CTS owing to osteosclerosis of cortical bone thus making the carpal tunnel smaller. Leri’s pleonosteosis is an extremely rare autosomal dominant disorder characterised by unusual facial features, short stature, skeletal malformations, limitation of joint movements, and abnormalities of the hands and feet. Median nerve compression occurs as a result of fibrocartilaginous changes in the flexor retinaculum and enlargement of the bases of the metacarpals (2, 25, 30).

Dejerine-Sottas syndrome is an autosomal dominant hypertrophic neuropathy in which multiple nerves are enlarged. Wasting of the muscle tissue and absence of reflexes are other characteristic features (2).

Weil-Marchesani syndrome is an autosomal recessive disorder of connective tissue, which presents with short stature, ocular problems, and brachydactyly with joint stiffness. A thickened flexor retinaculum can cause CTS (2).

Next to the causes above, we have the non-genetic causes of CTS. First of all, there are several idiopathic cases of CTS with childhood onset. These children have neither genetic nor systemic associated disease, no family history of CTS, and no local cause of CTS. One could think symptoms in idiopathic CTS in children would resemble those in adults. However their history and physical examination often differs from those in adults. Children tend to have symptoms for a long period of time before diagnosis is established and treatment instituted (25).

A typical case was described by Sieratzki et al (33): an 8-year-old girl was referred with long-standing problems of clumsiness, grip weakness and decreased growth of her right hand. By the age of three, her parents noted that the child often held the handlebars of her tricycle only with her left hand. By the age of six, she complained of occasional numbness and stiffness with pain in the right-sided fingertips. On examination the right hand was slightly smaller in size and there was obvious thenar wasting. Grip strength was weaker on the right side and two-point-discrimination in right fore- and middle finger was reduced, at 10 mm.

When open carpal tunnel release is performed, a thickened transverse carpal ligament is often found in children with idiopathic CTS, like in the patient reported by Wilson and Büchler. Open CT release showed the transverse carpal ligament to be extremely thick (4-5 mm). The median nerve had a dramatic hourglass deformity and appeared flattened and very pale (39).

Idiopathic CTS without thickening of the transverse carpal ligament has however also been described (25).

CTS in children caused by intensive sports practice has also been reported. According to Lamberti and Light (25) three sports have been reported as inciting CTS: golf, weight lifting and basketball. In the reviewed literature we came across six cases of CTS due to sports practice. One of them had complaints of progressive painful numbness over the first, second and third left digits after intensive basketball training, which persisted throughout the night (10). A second reviewed case had complaints of CTS related to skiing (12).
Tropet et al (36) reported four cases of CTS in children whose complaints arose during the day while normal activity was important and especially during sports activities like biking, playing tennis and rowing.

Several local aetiologies can be responsible for a carpal tunnel syndrome with childhood onset.

Chen et al (8) described nerve territory oriented macrodactyly (NTOM) as an unusual cause of CTS. NTOM is a benign form of localised gigantism. This is probably the same entity as simple fibrolipomatous hamartoma with the exception that there is also an increase in size of the bony and soft tissue structures of the involved digit or digits. The histogenesis is believed to be due to proliferation of fat and fibrous tissue within the nerve sheath or hypertrophy of mature fat cells and fibroblasts that are part of the epineurium. The pathologic changes of the nerves have been found to be identical whether they occurred with or without macrodactyly.

Salon et al (31) studied eight children with CTS due to fibrolipomas of the median nerve, of whom two had real gigantism of the involved digits. They also do not agree with the distinction between macrodystrophia lipomatosa, responsible for macrodactyly, and fibrolipoma of the median nerve. At surgical exploration, enlargement of the median nerve was noted in all reported cases. In most cases the enlargement had the aspect of a fatty infiltration, while in some cases the nerve had a more fibrous aspect.

Another tumour associated with CTS in childhood is intraneural perineuroma as described in a 2-year-old child by Alfonso et al (1). Intraneural perineuroma represents a tumorous proliferation of perineural cells, which diffusely infiltrates the endoneurium. Clinically it usually presents with motor deficit. Paresthesias may occur in some patients but pain is a rare finding, and it was probably due to the severe compression within the carpal tunnel in the above-mentioned case. Surgical exploration revealed an enlarged and hyperaemic median nerve and an intraneural tumour just distal to the origin of the palmar cutaneous branch. Microsurgical neurolysis revealed a dense fibrous intraneural lesion. The tumour was excised and the defect reconstructed with reversed sural nerve grafts.

A few cases of CTS due to a haemangioma of the median nerve have been described as well. In most cases the diagnosis was not suspected preoperatively as there were no associated cutaneous vascular lesions. In a case presented by Coessens et al (9) a 12-year-old boy complained of numbness and pain in the median nerve territory of the right hand. Clinical examination revealed a blue discolouration of the pulp of the index and middle finger and a soft tumour on the palmar aspect of the second web space. Exploration revealed a haemangioma around and within the median nerve, apparently arising from the vasa nervorum. Resection required extensive interfascicular dissection.

Children with haemophilia have been reported with spontaneous acute CTS secondary to local bleeding (25).

Another local cause of CTS is a musculotendinous malformation. Brax et al (7) divide these malformations into two groups: anomalies concerning the “regular” muscles and those concerning the “supernumerary” muscles. They describe six muscles that can be responsible. The palmaris longus muscle may be hypertrophic, inversed (tendon proximal and muscle belly distal), digastric, doubled or bifid (proximal or distal part double). The flexor digitorum superficialis muscle may have a prolonged muscular belly or a supplementary muscular belly or a digastric one. The flexor carpi radialis brevis muscle is a supernumerary muscle. The first lumbricalis muscle may have an insertion very high on the tendon of the deep flexor. There may be a tendinous binding between the deep flexor and the flexor indicis proprius. The palmaris brevis muscle can also cause CTS.

CTS has also been associated with a trigger finger. A 14-year-old girl presented by Cruz Martinez and Arpa had typical symptoms, i.e. painful paresthesia (10).

The cause of CTS can be a stenosing synovitis, tendon degeneration or muscular malformation (7). We would like to remark that in the survey of Poilvache et al (30), seven out of 52 patients with CTS (13%) had CTS associated with a trigger finger, while we could only retrieve one such case out
of the 163 cases we reviewed (when we do not consider the cases with a trigger finger associated with a storage disease).

The Klippel-Trénaunay syndrome (KTS) has been reported by Gassmann and Mettler (14) to cause CTS in a 12-year-old girl. The triad, first described by the French physicians Klippel and Trénaunay in 1900, consists of capillary malformations, varicosities and hypertrophy of bony and soft tissues. The aetiology remains obscure. Basherville et al (4) suggested that KTS is caused by a mesodermal abnormality during foetal development, leading to maintenance of microscopic arteriovenous communications in the limb.

In the case described by Gassmann and Mettler (14), CTS was caused mechanically by the capillary malformations and also by the altered haemodynamics that can cause intermittent lesser blood flow to the median nerve.

Poland’s syndrome is a congenital – mostly sporadic – anomaly classically consisting of the combination of unilateral aplasia of the sternocostal head of the pectoralis major muscle and an ipsilateral hypoplastic hand with simple syndactyly and short fingers. In the reviewed literature there were two cases of CTS caused by this syndrome, which is probably caused by a vascular disruption sequence of the subclavian arteries. CTS is probably caused by compression of the median nerve in the narrow carpal tunnel (7, 20).

One of the seven cases with childhood CTS described by Deymeer and Jones (12) had CTS secondary to scleroderma, a chronic autoimmune disease of the connective tissue. The 15-year-old girl had pain in the hand and difficulty in writing, but did not have typical CTS-symptoms.

Last but definitely not least, several cases of CTS in childhood due to trauma have been reported. In seven out of ten cases, the trauma caused epiphysiolysis of the distal radius (one Salter-Harris type I, six Salter-Harris type II).

Binfield et al (5) described three boys with displaced Salter-Harris type II fractures complicated by an acute median nerve compression. All three patients had paresthesias and sensory loss in the distribution of the median nerve, which developed within one hour of injury and subsequently increased. In each case the patient underwent immediate manipulation under anaesthetic and immobilisation in a split below-elbow cast with the wrist in slight flexion. Return of sensation and loss of paresthesias was immediate in two patients and within 24 hours in the third. Exploration of the median nerve was not required.

Deymeer and Jones (12) however suggest that early decompression may be indicated in traumatic median neurapraxias in children with potential for nerve entrapment. Nevertheless, of the two children with CTS after trauma they described, one was relieved of his symptoms without surgery and the other had an associated laceration of the median nerve, which did not recover.

Another case, not similar to the previous ones, has been reported by Brax et al (7). A 12-year-old girl developed compression of the median nerve six months after a Salter-Harris type I fracture. Compression was due to hypertrophic callus formation. Nerve decompression brought relief. The same authors report an 8-year-old boy with an ulnar and carpal tunnel syndrome appearing one month after distal fracture of the ulna and radius. Recovery occurred after section of the annular ligament, opening of Guyon’s canal and neurolysis of the two trunks.

Hodgkinson and Evans (22) also described two cases of post-traumatic CTS. The first one was a 9-year-old boy who lacerated his left wrist on a broken glass with division of the ulnar nerve and artery and of the tendon of flexor carpi ulnaris. Two weeks after surgical repair the boy complained of numbness and sensory loss and increasing pain. He was relieved of his symptoms after open release. The second one was a 14-year-old boy with pain in the right forearm, wrist and hand, 48 hours after his arm had been trapped between his thigh and the steering wheel of a cart while go-karting. On exploration the median nerve was found to be compressed by a fibrous band beneath the arch of the flexor digitorum superficialis muscle. This band was divided and complete motor and sensory recovery took place.

When we consider the treatment for CTS in children, most cases in the literature underwent open carpal tunnel release: this was the case for
145 of the 163 reviewed cases. While 141 cases improved after surgery, 4 needed a revision. One patient needed a second revision: the surgeons used a “turnover” palmaris brevis flap in conjunction with internal neurolysis (23).

CONCLUSIONS

Carpal tunnel syndrome during childhood is rare and often has an unusual presentation. Children often have modest complaints such as clumsiness and/or weakness, wrist or hand pain, thenar atrophy (differential diagnosis should be made with congenital thenar atrophy) and thenar weakness. Moreover, children often are too young to communicate their problem. In addition, childhood CTS usually occurs in specific aetiologies, such as storage diseases. In these cases the symptoms of CTS can be masked by the skeletal dysplasia and joint stiffness and some children may be unable to express their symptoms because of mental retardation.

We think it is clear that every child with even mild symptoms should be well examined and a thorough family history should be taken. Children with storage diseases may benefit from early clinical and electrophysiological screening before they develop obvious clinical signs.

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