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REVIEW PAPER

Peripheral nerve injuries in the pediatric population: a review of the literature. Part II: entrapment neuropathies

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Abstract

Introduction Entrapment neuropathies are infrequent in children, and therefore remain unrecognized. The incidence of radial, median, and cubital mononeuropathies are all similar. Despite the rarity of such cases, extensive, albeit scattered, literature has accumulated concerning entrapment neuropathies in children.

Objective To the literature concerning entrapment neuropathies in children.

Methods A systematic review of the existing literature has been made.

Results The management of chronic pediatric pain is very important in such patients to prevent youths from experiencing prolonged absences from school, sports, or other productive activities, and limit the psychological burden of chronic disease. Nonsurgical treatment of both cubital and carpal tunnel syndromes has been disappointing in pediatric patients, with only limited success; and, to date, there is no clear explanation for the outcome differences generated by nonsurgical management between adults and youths. Simple decompression of the ulnar nerve at the elbow also has much higher rates of failure in children than in adults.

Conclusions The presence of an entrapment neuropathy (specially carpal tunnel syndrome) in a pediatric-age patient should alert medical care providers to the potential of some underlying genetic condition or syndrome.

Keywords Entrapment neuropathies · Carpal tunnel · Ulnar nerve compression · Pediatric population

Introduction

Mononeuropathies are infrequent in children, with the incidence of radial, median, and cubital mononeuropathies that are all similar [1]. While direct trauma to nerves is not all that uncommon in children, entrapment neuropathies are considered exceedingly rare and infrequently recognized [2]. Despite the rarity of such cases, extensive, albeit scattered, literature has accumulated concerning entrapment neuropathies in children. This review analyzes this literature.

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Carpal tunnel syndrome

Idiopathic carpal tunnel syndrome (CTS) is uncommon in children, accounting for fewer than 0.2% of CTS cases; and the majority of cases are linked to genetic conditions [3]. One quarter of CTS cases in children are considered idiopathic.

One half of the cases of CTS are secondary to one of the lysosomal storage diseases, a group of inherited disorders that includes mucopolysaccharidosis and mucolipidosis [4]. CTS is a relatively rare complication in children with mucopolysaccharidosis types I, II, and III (e.g., Hunter's and Hurler's syndromes), with mucolipidosis patients most commonly manifesting CTS during childhood [5]. Median nerves in these storage diseases do not seem to be profoundly involved; the axons contain no abnormal deposits, and the nerve's Schwann cells and connective tissue are only mildly affected. It is the adjacent tenosynovium that is swollen, along with the tendons themselves; and this causes the CTS. The transverse carpal ligament is consistently thickened to 4–6 mm in these patients, versus the normal thickness of just 2–3.6 mm in non-affected adults [4]. The diagnosis of CTS is often delayed in children with lysosomal storage diseases,

because communication difficulties with young and often mentally challenged children renders the clinical presentation difficult to interpret. Treatment of the underlying metabolic disorder does not necessarily reverse CTS symptoms, so prompt surgical release often is necessary. Indeed, carpal tunnel release is recommended as soon as the diagnosis is established [6–8]. Children with a storage disease may benefit from early clinical and electrophysiological screening before they develop obvious clinical signs of CTS [9].

Other uncommon etiologies and unusual susceptibilities to CTS include congenital bone and muscle anomalies [10], hereditary neuropathy with liability to pressure palsies (HNPP), treatment with growth hormone, hypothyroidism, Down syndrome [11], hemophilia with localized bleeding in the region of the carpal tunnel, Schwartz-Jampel syndrome, scleroderma, multiple xanthomas associated with familial hypercholesterolemia, fibrolipomas, and fibrolipomatous hamartoma of the median nerves related to macrodactyly, obesity, rubella, and Klippel-Trénaunay syndrome [3, 9, 12–15].

The presentation of CTS in pediatric-age patients usually differs from the usual adult presentation. Hand clumsiness and thenar atrophy, rather than sensory complaints (tingling and pain) are often the presenting symptoms in children [5]. Since children with CTS often have modest complaints, despite long-standing difficulties like decreased manual dexterity in one hand or poorly localized pain, the diagnosis of CTS often is made late in the course of nerve compression [4].

In other, less-frequent cases, hand pain is the main symptom, associated with numbness and paresthesias, or night pains and underutilization of the first three fingers in the hand [16]. Hand or wrist pain are more frequently described in idiopathic and familial cases [4]. Adult-like presentations, with paresthesias and nocturnal symptoms, are more common in older children.

Physical examination reveals evidence of thenar atrophy in more than 50% of patients. In some cases, the degree of atrophy is so severe that it suggests the possibility of congenital absence or hypoplasia of the thenar muscles [4].

Provocative tests, like Tinel's and Phalen's test, are often normal in children with long-standing nerve compression. Electrodiagnostic studies are essential to establishing the diagnosis in many children [4].

The diagnosis is especially difficult in neonates and young infants, and usually made indirectly. Carpal tunnel syndrome has been reported in a 9-month-old infant who presented with intermittent abnormal movements of both hands [17]. Michaud et al. reported a patient in whom an unusual tendency to grasp with the fourth and fifth fingers was noted at 15 months [18]. Swoboda et al. reported a 7-month-old infant who presented with recurrent chewing of his digits in a median nerve distribution as the primary manifestation of carpal tunnel syndrome, in conjunction with clinical evidence of diffuse pain insensitivity. Electromyography (EMG) demonstrated severe median nerve entrapment at the wrist bilaterally. Carpal tunnel release resulted in complete clinical resolution and significant EMG improvement, and the child's self-mutilation was completely abolished [19].

This being said, the most frequent age for CTS diagnosis and treatment in children is between 6 and 8-years old [4].

Familial forms are usually dominantly inherited and only rarely present under the age of 5 years [20]. In such children, there often is a family history of CTS that does not skip generations. As stated earlier in this review, in such patients, symptoms are more typical of adult CTS, associated with pain, numbness, and nocturnal paresthesias [4]. Though the transverse carpal ligament has been described as enlarged in adults with familial CTS undergoing carpal tunnel release, this has not been observed in children with familial CTS [4, 20].

It has been reported that childhood CTS can be linked to certain sports (e.g., golf, weight lifting and basketball) and also to playing a musical instrument [21]. These patients typically are adolescents and can explain their symptoms more thoroughly, facilitating the diagnosis.

Bilateral presentation of CTS in children has also been reported [3, 17]. It is important to note that pediatric cases of CTS tend to demonstrate bilateral electrophysiologic abnormalities, even if only one hand is primarily involved clinically [4]. Therefore, it is recommended that EMG and nerve conduction studies (NCS) be performed bilaterally in all children suspected of having CTS.

Experience with pediatric CTS is limited. Conservative treatment of carpal tunnel syndrome (bracing, injection, etc.) in pediatric and adolescent patients has demonstrated only limited success, regardless of etiology. As such, it is not recommended [4]. Open surgical release is the only treatment documented to be effective in the treatment of childhood CTS and should be performed soon after electrodiagnostic confirmation [4, 22].

Outcomes of surgery for idiopathic CTS in children are poorer than for adults [23]. One proposed reason for this worsened prognosis is the protracted compression of the nerve that often occurs prior to the diagnosis being made [24].

Whenever CTS is diagnosed in a child, a thorough list of potential diagnoses should be constructed, because of the secondary nature of this syndrome in most pediatric patients. The scarcity of CTS in children and its frequently atypical symptoms may cause diagnostic delay, sometimes with serious consequences.

Proximal median nerve compression

Congenital constriction bands may cause median nerve entrapment, both proximally and distally, sometimes with concomitant radial and/or ulnar entrapment [21].

Proximal median nerve compression has rarely been reported in children. It has been reported and associated with

calcification of the flexor digitorum superficialis muscle [25], in individuals with the congenital anomaly called a "ligament of Struthers" [26], and in conjunction with a congenital supracondylar process [27].

Anterior interosseous nerve syndrome

Anterior interosseous nerve (AIN) syndrome consists of spontaneous pain on the ventral side of the elbow, associated with weak flexion of the terminal joints of the thumb and index fingers, resulting in a weak pincer grip.

This syndrome has been reported in several children, of either spontaneous onset [28, 29] or associated with a supracondylar fracture of the humerus [30]. Pronator syndrome has also been reported in children [31].

At surgery, abnormal fibrous bands, adhesions, or neuromas may be found; however, exploration often reveals no concrete lesion. Management is primarily conservative, with surgery indicated if spontaneous recovery is not evident after several months [32].

Ulnar neuropathy

Ulnar mononeuropathy is the most common upper extremity mononeuropathy in children [33], and its most common etiology is acute trauma.

Ulnar nerve entrapment can be associated with the syndrome hereditary neuropathy with liability to pressure palsies (HNPP), or with anomalous anatomy producing entrapment [5, 34].

Entrapment most commonly occurs in the cubital tunnel, but also may localize to the forearm, wrist, or hand.

Cubital tunnel syndrome in pediatric and adolescent patients is uncommon, relative to its incidence in adults [35]. Several studies have reported the occurrence of cubital tunnel syndrome in pediatric patients who participate in throwing activities [36–38].

Tardy ulnar nerve palsy associated with cubitus varus deformity after a supracondylar fracture of the humerus has been reported extensively [39–41]. The cause of ulnar nerve compression in these cases is kinking of the dislocated nerve at the proximal border of the flexor carpi ulnaris [39].

The symptoms of cubital tunnel syndrome are the same as in adults. They include elbow pain, numbness and tingling in the ring and small fingers, and intrinsic hand muscle weakness. On physical examination, the majority of affected extremities exhibit ulnar nerve subluxation at the elbow; while a positive Tinel's sign and positive elbow-flexion-compression test are each seen in approximately 50% of cases.

A distal compression injury of the deep ulnar branch at the wrist has also been described in an adolescent after prolonged bicycle riding [42]. This is a well-recognized mechanism in adults [32]. The presence of an epitrochleoanconeus muscle or congenital constriction bands has also caused ulnar entrapment in children [43, 44].

Nonsurgical treatment includes a trial of night-time splinting, activity modification, and anti-inflammatory medication. However, such conservative management is unlikely to relieve symptoms in this patient population. Stutz et al. reported an 83% failure rate with nonsurgical treatment [35]. Despite this high rate of failure, they recommended initial conservative treatment for cubital tunnel syndrome. They showed that surgical management can be efficacious and provides symptom relief, but also identified a surprisingly high failure rate of 23% with in situ decompression. The percentage of patients who require revision surgery also appears to be much higher than values previously published in the adult literature following simple open decompression [35]. Why the failure rate with simple decompression that is higher in younger patients is unclear. One explanation may be the different etiologies of cubital tunnel syndrome in children and adolescents versus adults. In adults, most cases are idiopathic and present with milder symptoms. This is unlike pediatric cases, which are more frequently associated with some congenital or posttraumatic etiology.

Suprascapular neuropathy

The suprascapular nerve is subject to damage from compression at the suprascapular or spinoglenoid notch. Entrapment of the suprascapular nerve is rare in younger children, but has been described in adolescents [45, 46].

The cardinal features of this entrapment are pain, posteriorly and laterally in the shoulder; wasting of the supraspinatus and infraspinatus muscles; and weakness of shoulder abduction and external rotation. In cases involving compression at the spinoglenoid notch, only the infraspinatus muscle is involved. It has been proposed that pain may be more prominent than weakness in younger patients [32].

Surgical decompression of the nerve usually results in early pain relief and good, though not always complete, restoration of strength.

Long thoracic neuropathy

The long thoracic nerve descends along the chest wall to innervate the serratus anterior muscle. Isolated palsies of the nerve are sometimes seen in childhood [32]. Long thoracic nerve compressions are clinically similar in children to what is commonly seen in adults. Causes of compressive neuropathy in children include spinal braces, prolonged pressure from carrying weights on the shoulder (e.g., a heavy backpack), and seatbelt compression [42, 47].

This neuropathy produces winging of the inner border of the scapula when the patient pushes against resistance, but not

at rest. Patients also are unable to raise their ipsilateral arm above their head [32].

It is frequently difficult to localize the point of compression. Fortunately, spontaneous recovery is the rule.

Radial neuropathy

Chronic radial neuropathies are rare in children; but can occur.

Pediatric compression-related radial nerve palsies most commonly occur in neonates. Feldman reported eight neonates with radial nerve palsy and attributed this to constriction of the fetal arm by abnormal uterine action [48]. Clinical observations support this hypothesis. Labor is often abnormally prolonged in such patients, particularly in the second stage, leading to potentially sustained intrauterine posture, wedging the arm against the pelvic brim to cause compression of the radial nerve. Sometimes, instrumental help (e.g., forceps) is required for delivery of the infant, which possibly also predisposes the nerve to compressive neuropathy [1]. In some of these patients, simultaneous subcutaneous fat necrosis or hematomas overlying the course of the radial nerve, proximal to the radial epicondyle of the humerus, has been described, which suggests the possibility of pressure against the nerve at this site. Associated skin necrosis also favors this entrapment mechanism [49].

Congenital constriction bands in the upper arm have been implicated in combined median, ulnar, and radial neuropathies, but not isolated radial nerve lesions [50].

Overall, the prognosis is excellent, with the majority of neonatal patients experiencing spontaneous full recovery within 5–6 months (mean 6–8 weeks) [1, 32]. However, neonatal radial palsies may be confused with a brachial plexopathy.

Escolar and Jones published a series of 16 pediatric patients with radial mononeuropathy. Eight (50%) of these cases were atraumatic, primarily related to compression and entrapment. The authors reported two additional cases of neonatal radial neuropathy related to prenatal intrauterine nerve compression and reviewed the literature, identifying a total of 32 cases. Six other cases they found were post-neonatal. These 38 neonates primarily had compression or entrapment of the radial nerve in the axilla or spiral groove, or of the posterior interosseous nerve in the forearm [1].

Radial nerve compression at the spiral groove, causing so-called Saturday night palsy, is a common mechanism in adults, accounting for 50% of radial nerve entrapment cases among adults; but this is rarely reported in children. As with neonatal cases, the prognosis in these children is excellent, with recovery expected within 6–8 weeks.

Entrapment of the radial nerve within the axilla also has been reported in children using crutches, who often present with bilateral radial palsy. Posterior interosseous compression also has been noted in child cancer patients secondary to an acute compartment syndrome caused by chemotherapeutic infiltration into the forearm [1].

These series all document how uncommon radial nerve entrapment is in children.

Very rarely, progressive radial neuropathies occur in children. These require a careful search for an entrapment site. In such patients, magnetic resonance imaging (MRI) can be of tremendous value identifying the anatomic site of compression. Any diagnostic delay may lead to irreparable damage of the nerve [1].

Progressive radial entrapment neuropathies also have been documented secondary to venipuncture-induced hematomas, entrapment within the lateral head of the triceps brachii muscle, and fibrous bands [51]. A radial nerve palsy also was reported in a preterm infant as the result of repeated blood pressure measurements [52]. Pediatric radial entrapment may also occur at the level of the posterior interosseous nerve from a tendinous process arising between the supinator and aponeurosis of the extensor digitorum communis [53, 54].

In any child with an evolving radial neuropathy, an EMG should be performed to confirm the neuropathy. An MRI also may be of value to localize the entrapment site and exclude other causes of entrapment (e.g., arcade of Frohse, lipomas, ganglia).

Musculocutaneous nerve

Musculocutaneous nerve compression is extremely rare in children and adults. One reported case was caused by compression secondary to wearing a body cast [47].

Posterior interosseous nerve

The posterior interosseous branch of the radial nerve passes through the supinator muscle and subsequently supplies the digits and wrist extensors. Its entrapment and compression may occur at the elbow and from fibrous bands or compartment syndromes in the upper forearm [32]. Similar to what is observed with other compression neuropathies in children, this is a very uncommon entity [55].

Ford described a 16-year-old girl with a long history of progressive painless paralysis of extension, initially involving the fingers and subsequently the wrist of the right arm, associated with progressive pronounced atrophy. Later, a similar condition developed in her left arm. Intra-operatively, the posterior interosseous nerve (PIN) was found to be greatly compressed between a tendinous process from the supinator muscle [54].

Meanwhile, Tubbs et al. reported on a 9-year-old boy with radial non-union and radial head instability that resulted in PIN compression due to recurrent dislocation of the radial head. Surgical decompression—which involved transecting the overlying supinator muscle and correction of the radial

deformity and instability—resulted in complete return of PIN function [55].

Thoracic outlet syndrome

Neurogenic thoracic outlet syndrome is extremely rare in children, though adolescents occasionally develop this unusual syndrome. The literature is largely comprised of single cases and small series [56].

The neurovascular bundle can be compressed by bony structures like the first rib, cervical ribs, and osseous tubercles, or by soft-tissue abnormalities like a fibrous band or muscle hypertrophy [57].

In adolescents, symptoms may be precipitated during rapid growth, since the anatomy of the thoracic apertures is constantly changing, especially in association with a cervical rib [58].

The symptomatology may be broad and relatively nonspecific; so the condition is easily overlooked [56]. Several presentations have been reported in childhood: recurrent torticollis [59]; local pain or a mass; vascular complications like peripheral gangrene and cerebral vascular accidents; intermittent aching pain along the ulnar side of the arm and forearm associated with sensory disturbances in a similar distribution; and upper limb weakness [32, 60]. Some young patients report that their affected arm "feels smaller" than their other arm [60]. Since children grow continuously, development of the affected limb can be affected as soon as the brachial plexus is compressed. Early decompression is very important in such cases [60].

The development of thoracic outlet syndrome has also been described in children secondary to surgical procedures for pectus excavatum [61].

Because so few cases of thoracic outlet syndrome have been reported in children and adolescents, specific therapeutic recommendations are lacking. A conservative approach to symptoms is warranted; first to allow further growth and remodeling of the thoracic outlet, which may be sufficient to accommodate the nerve roots and brachial plexus and alleviate symptoms [58]. On occasion, the presence of increasing or chronic symptoms may justify surgical treatment [62]. When motor deficits and atrophy are present (true thoracic outlet syndrome), decompression is indicated.

Common peroneal nerve entrapment

Compression of the common peroneal nerve at the fibular head is one of the most frequent forms of compressive neuropathy in childhood.

In neonates, peroneal palsy has been reported secondary to uterine bands, pressure from footboards or splints, and the infiltration of intravenous solutions [63–65]. In older children, causes are similar to those seen in adults: excessive pressure from an operating table during prolonged surgical procedures, compression from a splint or short leg cast, and prolonged crossing of the knees [66]. Bone tumors (osteochondroma) and exostoses have been reported as causes of compression of the nerve at the fibular head, so radiographic examination is necessary in children [67]. Spontaneous entrapment is not as frequent as in adults.

Children with peroneal neuropathy typically present with a unilateral foot drop. Both distal branches are involved in the majority of cases; less frequently, the deep branch is the only one involved [5, 66].

Factors predisposing to peroneal palsy include hereditary neuropathies like HNPP; significant weight loss in adolescence, sometimes due to anorexia nervosa [5, 66]; diabetes; and various forms of vasculitis, including anaphylactoid purpura [68].

The prognosis of compressive peroneal nerve neuropathy in children is usually good, especially with idiopathic compression, when spontaneous recovery usually occurs within a few months of symptom onset. Progressive lesions, and the absence of any spontaneous signs of recovery are indications to explore and decompress the nerve.

Sciatic neuropathy

Sciatic neuropathy is one of the most frequent neuropathies in childhood. In the past, a misplaced injection in the buttock region was the most common childhood cause of sciatic neuropathy [32], though this is much less common now. Nonetheless, the most frequent etiology remains traumatic.

Though compressive lesions of the sciatic nerve occur rarely in adults, in children they are not at all infrequent.

Prenatal compression of the sciatic nerve has been documented. Such prenatal injuries are thought to be secondary to external compression from reduced fetal activity, especially when associated with decreased amniotic fluid, amniotic bands, or uterine abnormalities [69]. In one series of 21 patients published in 2011, all the cases were associated with cesarean delivery, and the long-term prognosis generally was good [70].

The relatively undeveloped buttocks musculature of children offers the sciatic nerve little protection, which may account for their increased risk [47]. As such, the nerve may be compressed at the sciatic notch by prolonged sitting on hard surfaces or against one's heels [71]. Immobilization and prolonged unconsciousness in a supine position also increases the risk of sciatic nerve compressive injury in children [47, 72].

Entrapment of the sciatic nerve at the sciatic notch secondary to bony overgrowth of the posterior inferior iliac spine also has been described [73]. Venna et al. also described progressive sciatic palsy in a 12-year-old boy secondary to a constricting myofascial band in the lower thigh [32, 74].

Meralgia paresthetica

Meralgia paresthetica (MP) is a purely sensory mononeuropathy that is caused primarily by entrapment of the lateral femoral cutaneous nerve (LFCN) as the nerve passes deep to the inguinal ligament to supply the anterolateral region of the thigh. It has been well described in adults, but is very uncommon in children, with only isolated case reports in the pediatric literature.

In children, MP is typically idiopathic. However, some cases have been reported after posterior spinal fusion, with sports injuries, and with obesity [75]. Nowadays, the incidence of MP in the pediatric population could be exacerbated by rising rates of childhood obesity. Sanders et al. published a prospective study with the purpose of evaluating the incidence, risk factors, and time to resolution of MP after posterior spinal fusion for adolescent idiopathic scoliosis. They found that MP occurred in 25% of patients undergoing surgery; and this risk increased with longer operative times and heavier patient weight. On average, symptoms resolved in less than 4 days and there was no long-term pain or disability [76].

Typical symptoms are burning paresthesias and hyperpathia in the distribution of the nerve, which may be exacerbated by prolonged standing or walking.

The diagnosis is typically delayed in children, often leading to prolonged functional impairment and unnecessary medical testing [77]. Conservative treatment—including physical therapy, diet, and drugs (topiramate, gabapentin)—is the first step of management. An LFCN block confirms the diagnosis and provides analgesia.

Edelson and Stevens published a series of 20 children and adolescents with meralgia paresthetica. Ten of these patients had bilateral involvement. The average age at the onset of symptoms was 10 years (range, 1–17 years), and the average duration of symptoms before the patient was first seen was 24 months (range, 2–84 months). Twenty-four lesions were treated with open decompression of the LFCN, and 21 of these 24 patients were followed for at least 2 years. Fourteen of the 21 operations yielded an excellent outcome, with complete relief of pain and no activity restrictions; five led to a good outcome, with occasional pain, but no limitations in sports or other activities; and two resulted in a fair outcome, with pain that interfered with sports activities, but not with walking [78].

Meralgia paresthetica should be in the differential diagnosis for any child presenting with anterolateral thigh pain and no motor component. The condition is probably much more common in children than previously recognized [77].

Tarsal tunnel syndrome

Tarsal tunnel syndrome is classified as a focal compressive neuropathy of the posterior tibial nerve or one of its associated branches, either individually or collectively.

Tarsal tunnel syndrome is a rare condition, especially in the pediatric population, but can be debilitating if not addressed. It has been reported in association with mucolipidosis [79].

Few data exist in the literature on pediatric tarsal tunnel syndrome. In adults, the syndrome is equally distributed among the sexes, but almost all the children reported with tarsal tunnel syndrome have been girls [80].

The signs and symptoms are similar to those in adults [81]. However, Albrektsson et al. reported ten surgical cases of tarsal tunnel syndrome in children and highlighted how some symptoms are not usually seen in adults. One such symptom is recurrent sudden episodes of sharp pain in the foot. Six of their ten children walked with their affected foot in supination, putting weight only on the lateral border of the foot; while four had pain severe enough that it sometimes prevented them from putting any weight on that foot so that they needed to use crutches at intervals. All ten were operated upon, after which nine were symptom-free at follow-up and the tenth had improved [80]. Edwards et al. reported two patients who, since early childhood, had experienced difficulties with shoe fitting and preferred to walk barefoot.

Operative findings include local nerve swelling, a narrow tarsal tunnel, fibrous adhesions around the nerve, and constricting bands. The presence of an accessory muscle in the tarsal tunnel—the flexor digitorum accessorius longus or the abductor hallucis muscle—has been implicated as a cause of tarsal tunnel syndrome in children [81, 82]. The syndrome has also been reported to develop secondary to a previous tibial fracture.

Ultrasound-guided tibial nerve blocks have been used to relieve symptoms [83].

Surgery is indicated in younger patients, if conservative treatment fails. Surgical cases and small series have been reported in which very good results were achieved.

Hereditary neuropathy with liability to pressure palsies (HNPP)

This autosomal dominant condition presents as recurrent pressure palsies frequently related to trivial trauma and compression at common entrapment sites. A genetic basis was established in 1993, with most cases ascribed to deletion of chromosome 17p11.2-12 [84]. DNA analysis for this deletion is currently the major diagnostic criterion.

The age at which first symptoms first appear is within the first or the second decade in about 50% of patients, with clinical features apparent within 4 years. In some cases, palsies may be present at birth [32]. However, many patients are not diagnosed until much later, since the diagnosis is suspected only after recurrent episodes of neuropathy.

Patients develop single or multiple mononeuropathies, especially the peroneal nerve at the fibular head (35% of the cases), the ulnar nerve at the elbow (20%), the radial nerve

in the spiral groove of the humerus (9%), and the median nerve in the carpal tunnel (8%) [85]. However, any nerve may be affected, including the brachial plexus in 20% of patients [86] and the cranial nerves [32]. Proximal nerves tend not to be damaged in children with HNPP [86].

The pathology of HNPP is characterized by areas of focal thickening of the myelin sheath, known as tomacula [86].

Symptom onset is usually associated with trauma, which often is trivial, like sleeping on one's arm, sitting cross-legged, or resting on an elbow; however, many patients are unable to identify a precipitating traumatic event [87].

Known affected family members should be made aware of their predisposition to nerve injury and learn to avoid pressure damage. DNA analysis allows the easy detection of family members at risk, so preventative measures can be instituted early [32].

Conclusions

Entrapment neuropathies are very infrequent in children. Nonetheless, clinicians treating pediatric-age patients must always keep them in mind.

The management of chronic pediatric pain is very important in such patients to prevent youths from experiencing prolonged absences from school, sports, or other productive activities, and limit the psychological burden of chronic disease.

Some of these compressive neuropathies are potentially preventable. Physicians and nurses should be warned about the particular vulnerability of both the sciatic nerve to compressive injury in immobilized children, and of the peroneal nerve when children are positioned for operations.

Nonsurgical treatment of both cubital and carpal tunnel syndromes has been disappointing in pediatric patients, with only limited success; and, to date, there is no clear explanation for the outcome differences generated by nonsurgical management between adults and youths. Simple decompression of the ulnar nerve at the elbow also has much higher rates of failure in children than in adults.

The presence of carpal tunnel syndrome in a pediatric-age patient should alert medical care providers to the potential of some underlying genetic condition or syndrome.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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