REVIEW ARTICLE

Neuro-orthopaedic evaluation of children and adolescents: A simplified algorithm

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Summary Orthopaedic surgeons are often the first physicians to evaluate paediatric patients in the event of delayed walking, gait abnormalities, or parental concern about motor abilities. Therefore, orthopaedic surgeons must be thoroughly familiar with the normal neurodevelopmental stages. Neurological disorders are often first recognised during an orthopaedic evaluation. Minimal neurological abnormalities should be taken as warning signs that require additional investigations. Consequently, the evaluation must follow a strict protocol, even in children referred for apparently trivial functional disorders. We have developed an original physical examination protocol in which the largest possible number of signs is sought in each body position to ensure that the examination is both systematic and rapid. About ten minutes are required when all findings are normal. This protocol is extremely helpful for identifying the cause of the problem that motivated the evaluation or for reassuring the child and family. The main causes of paediatric orthopaedic disorders are cerebral palsy, spinal dysraphism, myopathies, peripheral neuropathies, motor neuron diseases, and intraspinal tumours. In some instances, no definitive diagnosis can be established clinically. In this situation, appropriate orthopaedic treatment can be initiated, although considerable caution is in order when establishing the indications. The cause may be detected only much later, when the clinical manifestations become more prominent.

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Minimal neurological abnormalities (claw toe deformity, mild muscle contracture, toe walking, or very brisk or absent reflexes) should be taken as warning signals. A previously missed neurological disorder is often recognized for the first time during a paediatric orthopaedic evaluation [1]. Consequently, a strict protocol must be followed when evaluating children, even those referred for apparently mundane functional disorders [1]. Here, we describe an exhaustive neuro-orthopaedic evaluation (Supplementary data, Appendix 1). The evaluation is standardised to ensure that no signs are missed. Neonates raise specific issues, which are not considered here. Our neuro-orthopaedic evaluation protocol is original in that each step seeks to detect the largest possible number of signs in a given body position (standing, walking, sitting, supine, and prone). This ensures that the evaluation is both thorough and rapid, with about 10 minutes being required when the findings are normal. Our protocol is extremely helpful for identifying the cause of the problem that motivated referral or for reassuring the child and family. Other evaluation checklists exist [2] but are more specifically intended for children with cerebral palsy (CP).

Physical neuro-orthopaedic evaluation

Observation of the child arriving in the examination room

The child should be observed for a limp (Trendelenburg gait due to gluteus muscle weakness or foot drop due to common peroneal nerve palsy), arm swing asymmetry (indicating mild hemiplegia), and other abnormalities. Attention should be given to upper limb use when sitting down, symmetry or asymmetry of the seated position, movements during undressing [3], and use of assistive devices (e.g., walking aids and orthoses).

Parent interview

The interview must be comprehensive, as opposed to focus on the reason for referral. The parents should be asked about age at walking, an attention deficit, difficulties at school (which may suggest visuoconstructive dyspraxia or dyslexia), and impaired sphincter function (suggesting spinal dysraphism). The interview often provides diagnostic orientation before the physical examination. The child’s health records should be checked for prenatal or postnatal problems and familial disease.

Observation and physical examination

The child should be fully undressed. Inspection is the first step: look for café-au-lait spots (neurofibromatosis), a midline lumbosacral marker of spinal dysraphism (lipoma, tuft of hair, angioma, or sacral dimple), calf muscle wasting (Charcot-Marie-Tooth disease) or hypertrophy (some forms of myopathy), lack of facial expression and ptosis (congenital myotonic dystrophy), and abnormalities of the hair and nails [4]. A standardised examination protocol enables monitoring of changes over time and facilitates communication with other healthcare teams. Muscle tone should be assessed and muscle weakness measured (Supplementary data, Appendix 2). The positions used to measure muscle strength should be varied to avoid exacerbating or diminishing synergies between muscle groups. If present, spasticity has no correlation with muscle weakness but may mask muscle weakness. As with joint angle measurements, the margins of error are wide, with considerable intra-observer and inter-observer variability.

Standing position

Evaluate the carriage of the trunk and head. Gluteus maximus paralysis results in an alternation of anterior and posterior trunk bending [4]. Exaggerated lumbar lordosis suggests weakness of the hip extensors or contracture of the hip flexors (CP or myopathy), whereas lumbar kyphosis is consistent with hamstring muscle contracture and global kyphosis with hypotonia. Fixed knee flexion may indicate quadriceps weakness or hamstring muscle contracture but may also compensate for pes calcaneus, pes equinus, or fixed hip flexion [1]. Malalignment or torsional deformity of the lower limbs should be sought. Determine whether the child places his or her feet flat on the ground [3]. In some children, excessive knee hyperextension may mask a pes equinus deformity (Fig. 1). Valgus of the hindfoot with pronation of the forefoot suggests CP, particularly when contractures are found at other sites. Acquired hindfoot varus is consistent with Charcot-Marie-Tooth disease, with supporting arguments being calf muscle wasting and even minimal wasting of the thenar muscles [1]; alternative diagnoses are hereditary degenerative diseases (Friedreich’s ataxia), myopathy, and spinal dysraphism [3]. Unilateral pes cavus suggests spinal dysraphism [4].

Ask the patient to bend and look for a gibbus deformity. Care should be taken to distinguish this finding from pseudo-gibbus deformity caused by leg length inequality, which resolves after compensation. The hand-floor distance provides information on whether hamstring contracture is present. Segmental stiffness of the spine should suggest a spinal tumour. Spine erector muscle function can be evaluated by having the patient bend backward and to each side. Asking the child to squat then stand up while keeping the arms folded assesses the flexion range of the three lower limb joints and the strength of the triceps surae, quadriceps, and gluteus muscles. A need to place the hands on the thighs to stand up from the squatting position (Gower’s sign) is an indicator of proximal lower limb muscle weakness, which is seen in all neuromuscular diseases; this sign is not pathognomonic for Duchenne muscular dystrophy.

The single-leg stance is useful to assess balance. Ask the child to hop on the toes of one foot while observing the ease with which the manoeuvre is carried out, assessing the shock-absorbing function of the foot, and looking for dynamic malalignment. Comparing the two sides during the monopodal hopping manoeuvre may provide valuable information. If asymmetry is detected, the child should be asked about any perceived difference between the two sides in terms of balance, concentration, or need for a greater effort on one side than on the other [3].

Evaluate the initiation of walking and any slowness in responding to the command. Check the three planes of space, the type of gait, its variability, the degree of


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synchrony between the upper and lower limb, upper limb swing, balance, foot and lower limb position before the attack phase, whether hip extension is preserved, whether knee flexion is inadequate or excessive, and knee orientation in the horizontal plane [5]. With experience, a gait abnormality can be rapidly detected, although a detailed analysis is difficult to achieve by physical examination alone. Dynamic equinus results in toe walking, particularly during brisk walking, whereas the heel is in contact with the floor in the standing position. Equinus deformity due to muscle contractures may cause either toe walking with flexion of the knee and hip or excessive knee hyperextension during the stance phase (Fig. 1). A digital camera can be used to film the patient walking back and forth twice, once face on and once from the side. Later on, the film can be used for a detailed analysis of gait.

Ask the patient to walk on tiptoes (triceps surae muscle weakness) and on the heels (tibialis anterior muscle weakness). During heel walking in a child whose feet are flat on the floor when standing, internal pes cavus with toe hyperextension and relative ankle equinus or knee hyperextension indicates paralysis or weakness of the foot interossei and lumbilcal muscles, suggesting Charcot-Marie-Tooth disease [3]. Hallux valgus deformity should be looked for.

Having the child move forward on the knees provides information on the gluteus maximus. A vertical position of the trunk indicates good gluteus maximus strength [4]. At the end of the visit, the child’s gait when leaving the room is more spontaneous and should therefore be carefully observed [5].

Supine position

Measure and record the range of motion of each joint. In children with CP, spasticity results in low reproducibility of motion range measurements [5,6], with variations of ±10% for same-day measurements and up to ±25% for measurements done several days apart [7,8]. Testing for spasticity is the cornerstone of the orthopaedic evaluation and cannot be separated from the evaluation of joint motion range [4]. The relative impacts of muscle length and muscle spasticity must be distinguished. The rapid angle reflects spasticity and the slow angle the degree of muscle contracture, if present, in patients with motion range limitation. Start at the ankle, where spasticity is easiest to detect. The Ashworth spasticity scale is widely used (Supplementary data, Appendix 3) [9]. Drawbacks of this scale include the absence of quantification of stage 0, the non-linear nature of passage from one stage to the next and, above all, failure to distinguish between functional impairment (true spasticity) and alterations in muscle mechanical capabilities (contracture) [6]. In patients with joint motion range limitation, the Tardieu scale [10] (Supplementary data, Appendix 4) differentiates the respective effects of muscle length abnormalities (via slow passive joint mobilisation) and spasticity (which occurs during rapid passive mobilisation). The Tardieu scale is more sensitive than the Ashworth scale [9]. Both scales should be used routinely to optimally evaluate spasticity. Co-contraction of antagonist muscles can be detected by abruptly changing the direction of the movement, which results in resistance. Resistance that appears immediately indicates co-contraction of antagonist muscles [6]. Evaluate the child for motor or sensory impairments consistent with a spinal tumour. Absence of the abdominal cutaneous reflexes suggests syringomyelia. The child should be asked to sit as a test of abdominal muscle strength. Ankle and feet. Calf circumference measurement on both sides is helpful to detect muscle wasting or hypertrophy. The two-step Silfverskiold test is used to assess ankle dorsiflexion (Fig. 2). Soleus muscle contracture is detected with the hips and knees flexed at 90°. The angle between the sole of the foot and the axis of the leg is recorded. While holding the foot to maintain dorsiflexion of the ankle, the knee is gradually extended to look for a decrease in the initial angle related to involvement of the entire triceps surae (soleus and gastrocnemius muscles) (Supplementary data,
Failure to evaluate triceps surae tone during the initial orthopaedic evaluation precludes the detection of non-articular musculotendinous contracture (CP). Several passive test results in combined ankle clonus (Supplementary data, video 3). Plantar flexion is assessed with the knee flexed. Measure the range of motion of the subtalar and midtarsal joints, assess the ankle tendon reflex, and look for a Babinski sign. Hallux valgus deformity in a young child often indicates an architectural abnormality of the hindfoot. Isolated weakness of the extensor hallucis longus muscle suggests early-stage peripheral neuropathy [4].

Knee. The popliteal angle is measured by starting with the hip and knee flexed at 90° then maximally extending the knee (Supplementary data, videos 4 and 5). Rapid knee extension results in resistance due to the stretch reflex, which reflects spasticity. When the knee extension is then continued slowly (slow angle), the degree of muscle and tendon contracture can be measured. The angle is positive when the axis of the leg is above the horizontal plane and negative otherwise. Thus, an angle of 120° between the thigh and leg notch is noted as 90° + 30°. Beyond 180°, the angle of fixed knee hyperextension is measured, and below 180° the angle of fixed flexion. Unilateral popliteal angle measurement is performed with the other hip extended and bilaterally measurement with the other hip flexed to maintain pelvic alignment. The bilateral angle exhibits better intraobserver and inter-observer reproducibility and is the only angle correlated with hamstring length as evaluated during gait analysis [2]. Caution is in order when only static information is available, and popliteal angle measurement alone is not sufficient to determine that operative hamstring lengthening is in order [11]. Knee flexion is assessed with the hip flexed to 90°, by measuring the angle between the axis of the thigh and the axis of the leg. Improvement of knee flexion upon flexion of the hip indicates contracture of the rectus femoris muscle. The heel-buttock distance can be measured. Patella alta is assessed by measuring the distance between the tip of the patella and the edge of the tibial plateau identified by palpation, with the knee flexed to 45° [6]. Finally, ligament stability should be assessed, patellar grating looked for, and the straight leg-raising test performed.

Hip. First, measure active (Supplementary data, video 6) and passive (Supplementary data, video 7) flexion of the hip. Fixed flexion deformity is detected by flexing the other hip until physiological lumbar lordosis occurs. Do not continue until kyphosis occurs, which would continue in the angle of fixed flexion. In general, the contralateral hip is flexed to about 110°, which corrects the exaggerated lordosis induced by any limitation in extension of the examined hip. Estimating “physiological lumbar lordosis” is subjective and, therefore, dependent on the examiner. Measure the angle between the spine and the table. Unresponsiveness of the fixed hip flexion to flexion of the knee indicates shortening of the psoas muscle, whereas an increase in hip flexion upon flexing the knee indicates rectus femoris muscle contracture (Thomas test) (Fig. 3). Two maneuvers are used to test hip abduction: the monoarticular adductor muscles are tested with the hips extended and the knees flexed (Supplementary data, video 8), whereas the bi-articular gracilis muscle is tested with the hips and knees extended (Supplementary data, video 9). When the hips and knees are flexed, abduction should be symmetrical and at least equal to 45° on each side. A smaller angle or asymmetrical hip abduction requires evaluation by a pelvic radiograph. The child must be completely undressed to enable detection of a pelvic tilt by inspection of the intergluteal cleft [4] (Fig. 4). Hip abduction angles may be affected by excessive antetorsion [2]. Fixed hip abduction (due to contracture of the gluteus medius and tensor fascia lata muscles) cannot be distinguished from fixed hip flexion (indicating contracture of the direct flexor muscles, i.e., the rectus femoris, psoas, and sartorius) if the patient is examined only in the supine position, as both abnormalities are associated with exaggerated lumbar lordosis that resolves when the hips and knees are flexed [4]. An additional evaluation in the prone position is necessary. Adduction should be tested with the hips extended and may be limited by contraction of the abductor muscles. To assess the gluteus maximus muscles, both with the patient’s lower limbs extended, the examiner can lift both heels about 10 cm above the table and ask the patient to lift the pelvis in the bridge position (Supplementary data, video 10). By releasing the heels one after the other, asymmetry can be detected. Spasticity of the gluteus minimus can be detected based on a catching sensation during external rotation. The circumference of both thighs should be measured to look for muscle wasting or hypertrophy.

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**Upper limbs.** Look for motion range limitation of the shoulders, particularly in external rotation; decreased elbow extension; forearm pronation; and more or less reducible flexion of the wrist. Thumb-index pinch grip and active thumb abduction should be assessed [4]. Evaluate the patient for spasticity, measure muscle strength, and record any muscle wasting (thenar eminence). Constitutional laxity of the thumb may suggest Marfan syndrome or Ehlers-Danlos syndrome [4]. Finally, the reflexes should be tested.

**Prone position**

With the patient in the prone position, assess passive (Supplementary data, video 11) and active (Supplementary data, video 12) knee flexion. Examination in the prone position is crucial to differentiate fixed hip abduction and fixed hip flexion. A limited range of abduction with persistence of the lumbar lordosis when the lower limbs are spread apart indicates pure flexion deformity. On the contrary, if abduction is possible over a wide range (often 90° on each side) and abolishes the lumbar lordosis, then the patient has pure abduction deformity. Unilateral abduction deformity results in tilting of the pelvis [4]. Concomitant flexion and abduction deformity is possible but exceedingly rare. Finally, hip rotation angles should be measured, with the knees flexed, which allows an evaluation of femoral antetorsion by palpation of the greater trochanter while moving the hip through its range of rotation (Netter’s method) (Supplementary data, video 13) [12]. Leg bone torsion is assessed with the knee flexed by measuring either the angle between the axis of the knee and the bimalleolar axis (anatomical torsion) or the angle between the projected axis of the femur and the line through the midfoot or medial edge of the foot (Supplementary data, video 14). The hindfoot should be held firmly to avoid spurious tibial torsion due to midfoot distortion. Hindfoot valgus/varus is measured first with the knee flexed then with the knee extended. Both adduction/abduction and pronation/supination of the forefoot should be determined. Examination of the foot is crucial,

**Figure 3** Thomas test used to measure contracture of the hip flexor muscles (psoas and rectus femoris): a: an increase in the angle between the thigh and the table when the other knee is flexed indicates a contracture predominantly affecting the rectus femoris muscle. Caution is in order, as a pure fixed abduction deformity can simulate a pseudo fixed flexion deformity. Having the patient hold the other thigh may substantially increase the spasticity. To avoid this phenomenon, the flexion of the contralateral thigh can be maintained by the examiner or another person; b: contracture of the rectus femoris muscle can also be suspected based on a decrease in knee flexion when the child is at the edge of the table.

**Figure 4** a: when the patient is wearing underpants or a drape, hip abduction seems roughly symmetrical; b: with the child fully undressed, the intergluteal cleft is seen to be deviated; c: when patient position is changed to place the intergluteal cleft in the vertical position, hip abduction is clearly asymmetrical.
Sitting position with the legs hanging over the edge of the table
The pectoralis major and latissimus dorsi muscles can be tested by lifting the child up from the arm pits. If this manoeuvre results in upwards displacement of the shoulders, the patient should be evaluated for hypotonia or paralysis of these two muscles. Assess the carriage of the head and trunk, as well as the patient’s ability to voluntarily straighten the spine in the sagittal plane. Backward bending of the trunk indicates either muscle weakness affecting the psoas and/or rectus abdominis or contracture of the hamstring muscles. Use by the child of the upper limbs when asked to sit up straight indicates weakness of the trunk muscles [4]. Note whether the pressure on the buttocks is symmetrical between the two sides (presence or absence of pelvic tilt). Look for gibbus deformity, evaluate passive rotations of the trunk, and push the child in the anterior-posterior and side-to-side directions to assess automatic balancing responses. The knee extensor lag angle is the difference between maximal active and passive extension of the knee (Supplementary data, video 20). Knee extensor lag indicates weakness of the quadriceps muscle (patella alta), excessive antagonism by the hamstring muscles, or both [4,6]. The planter flexors are tested against mild resistance (applied by the hand of the examiner placed flat under the sole of the foot) and the dorsiflexors against gravity (Supplementary data, video 21). Ankle motricity is difficult to assess in patients with spasticity and co-contractions. An underrecognised sign of minimal-symptom CP is involuntary tibialis anterior contraction synergistically with opposed flexion of the hip (Strümpell manoeuvre). This sign can be looked for starting at 4-5 years of age. The examiner asks the child to raise the thigh by actively flexing the hip while leaving the knee and ankle completely relaxed. When this manoeuvre is repeated while the examiner’s hand opposes the thigh elevation, involuntary active dorsiflexion of the foot occurs (Supplementary data, video 22) and the tibialis anterior tendon becomes visible under the skin [3]. Finally, malalignment of the patella in the last 20° of knee extension should be looked for and the patellar reflexes assessed.

The diagnosis is obvious or strongly suspected
This situation occurs with six main diagnoses.

Cerebral palsy
CP is a group of disorders affecting movement and/or posture and motor function. CP is estimated to occur in 1 to 2.5/1000 births [14]. The term CP applies to children and adults with motor impairments due to non-progressive brain lesions, regardless of their cognitive abilities and of the cause of the brain damage [4]. Most individuals with CP have a history of premature birth or perinatal distress. Although CP is a lifelong disorder, the clinical manifestations may change over time. Spasticity is a feature in 85% of cases, dyskinesia in 10% (with chorea, athetosis, and/or dystonia), and ataxia in 5%. Patients with ataxia have cerebellar dysfunction and impaired voluntary motor coordination [12]. Mixed forms of CP combine spasticity with dyskinesia or ataxia [12]. Depending on the site of the brain lesions,
the patient may have tetraparesis, diplegia, hemiplegia, or monoparesis. Delayed walking and pyramidal syndrome are consistent features. Muscle contractures related to spasticity result in skeletal deformities.

**Spinal dysraphism**

Spinal dysraphism (spina bifida) is a focal birth defect of the spine that occurs at the end of the first month of embryonic life. The posterior arches of the vertebrae fail to close. The defect is usually located at the lumbar spine or sacrum. Spina occulta is very common (10% of the general population) and usually asymptomatic, being diagnosed only radiographically. In spina aperta, the spinal cord and meningyes are herniated through the bony defect (myelomeningocele). Spina bifida is nearly always combined with other defects such as Arnold-Chiari malformation, hydrocephalus, and primary or secondary syringomyelia [15,16]. Less severe forms of spinal dysraphism include lipomas, diastematomyelia, and meningocele. The incidence of myelomeningocele has decreased as a result of advances in the prenatal diagnosis of this defect and of routine folic acid (vitamin B9) supplementation of pregnant women. Orthopaedic deformities develop chiefly in the hips, knees, feet, and spine. The main prognostic factor is the spinal level involved.

**Myopathies**

Duchenne muscular dystrophy is a progressive disorder inherited on an X-linked recessive basis. The incidence is 1/3500 boys. Gradually increasing muscle weakness and fibrous contractures of the muscles and fasciae result in skeletal deformities and severe functional impairments [17]. By 10 years of age the child is no longer able to walk, and severe spinal deformities develop in the second decade of life. The disease is fatal in the third decade, due to cardiorespiratory failure. The physical findings consist of hypotonia, mild motor development impairments, a slight delay in walking and, in some cases, toe walking. Hypertrophy of the calf muscles and exaggerated lumbar lordosis are apparent. Muscle strength is diminished, particularly at the proximal limbs. Given the predominant involvement of the gluteus muscles and hip extensors, climbing stairs is increasingly difficult and soon becomes impossible. Fatigability when walking is noted. When sitting on the floor, the child cannot stand up without pushing his hands on his knees (Gowers’ sign).

Congenital myotonic dystrophy (Steinert’s disease) is a muscle disease inherited on an autosomal dominant basis, with nearly complete penetrance and variable expressivity [18,19]. From one generation to the next the disease tends to develop earlier and to be more severe, a phenomenon known as anticipation.

**Peripheral neuropathies**

Charcot-Marie-Tooth disease (CMT) is a group of inherited chronic peripheral neuropathies characterised by degeneration of both motor and sensory nerve fibres [20,21]. Its prevalence is 1/2500 [20]. All patterns of inheritance are found: autosomal dominant in about 75% of cases, recessive, and X-linked (dominant or recessive, about 20% of cases). Some patients have sporadic disease [20]. Nerve fibre degeneration occurs gradually and progresses very slowly, causing muscle weakness that starts at the fibular muscles. The first symptoms usually occur at about 3 years of age, although late-onset forms exist. Walking may be delayed. About 50% of patients have gait disturbances with foot drop, a waddling gait, unsteadiness in the standing position, or difficulty running with frequent falls. Pes planovalgus predominates between 2 and 5 years of age, after which the deformity progresses to pes cavovarus with claw toe deformity and, in some cases, pes equinus. The impairments worsen slowly, with a faster pace of progression at puberty. Distal muscle wasting should be looked for at the feet and hands (atrophy of the thenar and hypothenar eminences and of the interosseous spaces, particularly between the first and second rays). Gradually, the disease progresses to the typical picture combining pes cavovarus with or without pes equinus, calf muscle wasting, wasting of the distal third of the thigh, and weakness of the small hand muscles with, in particular, thumb-index grip impairment. Over time, patients with Charcot-Marie-Tooth disease develop distal paralysis with muscle wasting and reflex abolition that predominantly affect the lower limbs. Thoracic kyphosis or scoliosis may result in severe deformities.

**Inherited degenerative spinocerebellar diseases**

The most common is Friedreich’s ataxia, an autosomal recessive disease due to a mutation in a single gene on chromosome 9. Its prevalence is 1/100 000 [22]. The neurological manifestations indicate involvement of the cerebellum, pyramidal tract, and dorsal column and eventually result in skeletal deformities [1]. Pierre Marie disease is responsible for severe ataxia. Strumpell-Lorrain disease, also known as hereditary spastic paraplegia, is inherited on an autosomal dominant basis and manifests as progressive spasmatic paraplegia with a cerebellar syndrome.

**Motor neuron diseases**

Spinal muscular atrophy (SMA) or Werdnig-Hoffman disease affects the neurons in the anterior horns of the spinal cord. The disease is inherited on an autosomal recessive basis and is due to a mutation on chromosome 5. Three clinical patterns have been differentiated [23,24]. Children with SMA have weakness of the trunk and limb muscles.

Polioyelitis is a viral disease that has been nearly completely eradicated in France via routine immunization. The disease affects the anterior horns of the spinal cord and therefore causes isolated motor impairment [1]. The site and extent of the spinal cord lesions vary widely across patients, resulting in a broad array of clinical pictures.

**Spinal tumours**

Intraspinal tumours are the most likely to cause neurological manifestations. Intraspinal tumours may be extradural...
(neuroblastoma, epidural metastases, hour-glass neurinoma), meningeal (meningioma), intradural (neurinomas), or located within the spinal cord (astrocytoma, ependymoma, dermoid cyst, teratoma, lipoma).

The diagnosis is not obvious

In some patients, no definitive diagnosis can be established. Evaluation by a paediatric neurologist and/or multidisciplinary team is extremely beneficial to the diagnostic process and helps to select the best investigations. Even when the diagnosis is not obvious, reasoning by elimination may narrow down the field of possibilities via appropriate investigations. The absence of a definitive diagnosis does not preclude the initiation of necessary orthopaedic treatments, although great caution is in order in the indications (e.g., do not perform extensive tenotomy if a neuromuscular disease is suspected). The cause may be identified only much later, when typical clinical manifestations develop. Strenuous efforts must be made to identify the cause, with the goal of selecting the optimal treatment strategy. In some cases, the diagnosis is established but the pattern of skeletal deformities over time is unexpected, either because unexplained relapses occur or because the course is atypical. In this situation, the case should be reappraised and perseverant effort on the part of the team may be the only means of correcting the initial diagnosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

Appendix A. Supplementary data


References