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PEDIATRIC ORTHOPAEDICS

REVIEW

CLINICAL CASES

HELLENIC ASSOCIATION OF ORTHOPAEDIC SURGERY AND TRAUMATOLOGY

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EDITORIAL

Paediatric Orthopaedics
A Chore or a Challenge?

John K. Dimitriou

Our specialty, Orthopaedics, as we all know, is the branch of medicine, of surgery in fact, which deals with the function of the musculoskeletal system and involves the prevention, treatment and restoration of congenital and acquired diseases and injuries.

For centuries, diseases of the musculoskeletal system, and especially injuries, were a "job" of the General Practitioner and the General Surgeon. Fractures, in particular, used to be the "territory of specialized Bone Setters". Gradually, and particularly the last two or three centuries, physicians started to focus their scientific interest on musculoskeletal system, thus "sowing the seeds" of Orthopaedics.

However, particular emphasis was given to the musculoskeletal system and its ailments by the father of medicine, Hippocrates, who, in its everlasting work, makes extensive reference to injuries as well to congenital abnormalities and other Orthopaedic conditions.

The first use of the term "Orthopaedics" was made over two and a half centuries ago, by Nicolas Andry in his classic textbook "Orthopedie" which was published in Paris (1741) and in London (1743). Thus our specialty gained a name and nowadays it can celebrate 275 years of official existence. From then, much time was to pass before our specialty escaped completely from the "embrace" of General Surgery and with the help of the technological advancement, progressed by leaps and bounds to the point which we all recognize today.

But this same specialty of Orthopaedics, like every young sapling, developed, sprouted and acquired many branches and twigs which gradually began to separate into subspecialties. One of these is Paediatric Orthopaedics, which has already passed from its empirical form into scientific theory. One might justifiably wonder whether this "fragmentation" of our specialty, or of any specialty, is scientifically correct. Will we eventually go to greater extremes? Will we have, say in a few years, one surgeon for the right hand and another for the left? Certainly not!

As regards the area of Orthopaedics which relates to children, I would like to be permitted to raise the following simple arguments:

1 The organism of the infant and child is not that of a small adult, but has numerous and central peculiarities that, in my opinion make it quite distinct. The potential of dynamic development and growth differentiates it from the anatomical and physiological point of view. The presence of growth plates creates conditions which pose problems in the treatment of certain diseases and so excludes methods applied in adults.

2 The capability of rapid response in healing and the union of fractures is taken for granted in children, while the process of remodeling physiological function often leads to surprises, positive or negative.

3 The tolerance and resistance of the child's joints to immobilization is also taken for granted. Furthermore, the effect of heredity must be investigated by means of a discreet examination of the parents or, if this is difficult, through appropriate questions.

4 The manner and the behavior of the doctor towards the orthopaedically sick child, while obtaining the history, the in-hospital treatment, and the subsequent follow up, require special attention and ability, since we all know that parents and children often mislead us by focusing the symptoms on a point far away from the afflicted region.

EDITORIAL

5 "Informing the parents": In my opinion, this is one of the most important, perhaps the MOST important, of the doctor's duties and obligations. Without doubt, it demands time, patience and persistence.

Imagine a young couple who have just had their first child, who presents some skeletal deformity or congenital musculoskeletal malformation, or who after a difficult birth, has a brachial plexus injury.

6 One other particularly difficult mission for the Orthopaedic specialist involves the cases of musculoskeletal malignancies in children. In such cases, the doctor is expected to offer, above all, psychological support and encouragement to the anxious parents.

Of course, Paediatric Orthopaedics has not been formally recognized; however in the mind of the Orthopaedic Surgeon there is a tendency to refer paediatric orthopaedic cases to those more specialized.

The widening of the field of General Orthopaedics, in combination with technological progress and the scientific and civil responsibility of the Orthopaedic Surgeon, requires under present conditions that the trainee, in the course of his/her training should have at least some basic knowledge of the orthopaedic problems in children. And this is not so that he/she will be in a position to treat every paediatric orthopaedic condition, but so that he/she at least will be able to differentiate the pathological from the physiological in borderline cases. Thus he/she will be in a position to guide the parents appropriately, rather than leaving them in the distressing state of complete ignorance and under a psychological burden, especially in cases when they are unable to consult someone else or to refer the case to a specialist.

Let us imagine here a simple case of a pulled elbow, which might cause difficulties to someone who had no contact with Paediatric Orthopaedics in the course of his/her training. He/she perplexedly examines the small child, asks for X-rays of the elbow, shoulder, and so on, but is unable to relieve the child's suffering or to reassure the parents. Of course, he/she may be lucky in that, during the manipulations or automatically during the positioning for the X-rays, the condition might be resolved. But if this does not occur, and the parents seek the assistance of another Orthopaedic Surgeon who has served in a Paediatric Orthopaedics department, or who has at least seen such a case once before, then the well-known simple manipulation will free both, the child from the pain and the parents from their anxiety. And this will surely cause at least a "pang of conscience" to the first unfortunate physician.

Another factor which further reinforces the importance of this field is the publication of special medical journals on Paediatric Orthopaedics, the appearance of special chapters on the topic in older journals and, especially, the publication of textbooks devoted exclusively to Paediatric Orthopaedics.

At this point, we should note and reflect on the fact that the first textbook, by Tachdjian, which was written purely about Paediatric Orthopaedics was only published in 1971(!), while later, and in particular during the last ten years, there have been repeated editions of textbooks on General Paediatric Orthopaedics as well as monographs on special Paediatric Orthopaedic topics.

How, though, do we come to be talking about Paediatric Orthopaedics? The great revolution, the recognition of the right and the obligation of a group of colleagues to practice with special interest in the orthopaedic problems of childhood, came from America, led by William Green, who is considered to be the father of Paediatric Orthopaedics. So, in 1971, the Paediatric Orthopaedic Society of America was formed, consisting of 11 members. A little later, in 1974, another small group of American Orthopaedic Surgeons with common interests founded the P.O.S.G. (Paediatric Orthopaedic Study Group). These two groups united in 1983, to form the well-known P.O.S.N.A. (Paediatric Orthopaedic Society of North America), which gathered into its fold all the American Orthopaedic Surgeons who were involved with Paediatric Orthopaedics. Around the same time (1982) Europe too found its Paediatric Orthopaedic voice with the creation of the European Paediatric Orthopaedic Society, (E.P.O.S), of which I had the privilege to be among the first members.

In Greece, many years ago, those involved in Paediatric Orthopaedics formed a group and, in regular meetings in the Paediatric Hospitals, in Athens, in turn,

we had an exchange of views and discussions of cases, while at the same time we participated regularly in the official annual congresses of the E.P.O.S.

In 1989, the Section of Paediatric Orthopaedics was created within our big mother Society (E.E.X.O.T) and now has around 40 members from all over Greece. As I mentioned before, the Orthopaedic Surgeons who were involved in Paediatric Orthopaedics took these initiatives out of a need for scientific information and discussion. Events, however, have overtaken the status quo and have brought to light new difficulties and questions.

Should the aspiring Orthopaedic Surgeon come into contact with or practice in general or specialised Orthopaedics during his/her training and, if so, for how long? In such a case, would this be beneficial both for the doctor and patients? Is it a waste of time, or is it an opportunity for the trainee to get to know corners of his/her specialty which have both great scientific interest for him/her and essential significance for the orthopaedic well-being of children?

In practice, a universal system on the necessity and the length of time devoted in training in Paediatric Orthopaedics does not exist. In U.S.A the Fellowship in Paediatric Orthopaedics is obligatory for somebody wishing to practice Paediatric Orthopaedics. In Greece, training Paediatric Orthopaedics is not compulsory. Only ten to fifteen percent of the young trainees, of their own volition and on their own initiative, are formally trained for six to twelve months during their six years of specialization Twenty years ago, a new law of the Ministry of health was voted requiring one year in Paediatric Orthopaedics ("cold" and trauma), but NEVER was applied in practice.

In March 2015 E.F.O.R.T, (European Federation of Orthopaedics and Trauma) in which E.E.X.O.T (Hellenic Association of Orthopaedics and Traumatology), is a finding member published the so called European Curriculum in Orthopaedics and Trauma, as European Education Platform. Hoping that this will be the starting point of an acceptance of guides, at least for Europe!

As we can see, things have been straightened out, and currently it is an established necessity that the training in Paediatric Orthopaedics is compulsory for a period of six to twelve months.

Some years ago, I sent to all the Orthopaedic Clinics (University and State Clinics that are formally recognized for Orthopaedic training) throughout Greece, a questionnaire about their capabilities of dealing with Paediatric Orthopaedic cases and their thoughts on the necessity of formal Paediatric training during the specialization. Most replied, which showed the real anxiety and difficulties, and it was mainly those who do not have Paediatric Orthopaedic cover and are unable to cooperate with nearby special centers in their region (particular geography, with many islands and remote areas). They were unanimous in agreeing that trainees, in the course of their specialization, should receive formal training in Paediatric Orthopaedics too. Of course, nowadays the "new generation" of Doctors are more privileged, as they can easily access sources of Paediatric Orthopaedics, such as distance e-learning, special national or inter-national meetings, conferences etc. But allow me to say and remind what, our Professors and Mentors at the University used to say: "Medicine is learned at the side of the patient – child."

Finally, is Paediatric Orthopaedics a CHORE or a CHALLENGE?

I think we all agree that it is not a chore, but an essential component of the whole for every doctor who has chosen the Orthopaedic specialty. Is it though a challenge? I think that the answer to this question is positive and arises from, among others, the enthusiastic opinion of those who have approached it, have got to know it, as well as from the perplexity of those who have ignored it or, for reasons apart from their own choice, have not had the opportunity to encounter it.

However, it is never too late. Even now let there be a start, a leap forward, and from here on let us offer this challenge to the young future colleagues within our Specialty. The childhood population of Greece (0-14years) is estimated about 2.000.000. I think all these children deserve the best Orthopaedic health!

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REVIEW

Spinal Deformities in Neurofibromatosis Type 1

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ABSTRACT

Neurofibromatosis type 1 (NF-1) is the most common human single-gene disorder. Skeletal complications usually present early in life and can be attributed to abnormalities of bone growth, remodeling, and repair in NF-1 or can be secondary to nearby soft-tissue abnormalities associated with NF-1. Scoliosis is the most common osseous manifestation of NF-1. It is important to recognize the dystrophic curve and to distinguish it from the non-dystrophic curve. The management of spinal disorders in young children in NF-1 continues to be problematic. The use of growing rods allows more longitudinal growth than fusion and more life freedom than bracing. The problems we have encountered are mechanical and could be expected when proximal and distal fixation is performed over an otherwise completely mobile spinal column. The multiple surgeries increase the potential for complications including infections. We continue to pursue solutions to our problems. The intent of this article is to present the spinal deformities that are most commonly associated with NF-1 and to identify the current management of spinal disorders based on the most recent literature.

KEY WORDS: neurofibromatosis; scoliosis; kyphosis; dystrophic deformity; NF-1

1. Introduction

Neurofibromatosis is a multisystemic, autosomal dominant genetic disorder defined as a spectrum of multifaceted diseases involving neuroectoderm, mesoderm, and endoderm. The clinical features of neurofibromatosis type 1 (NF-1), the most common form of the disease, were reported in several family members by German pathologist Virchow in 1847 [1], but it was his student von Recklinghausen [2] who 35 years later described the histological features of the syndrome that often bears his eponym.

NF-1 is characterized by extreme variability of expression. The proposed mechanisms for this variability include germline-modifying genes, environmental agents, second hit somatic mutation events in NF - 1 or other genes, epigenetic modification, and post-zygotic mutations [3]. The NF-1 phenotypes vary to a greater degree with increasing dis-

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tance from a proband, thus documenting that the specific familial NF – 1 mutation is not the primary cause of variability [4]. Common clinical manifestations include café-au-lait macules, neurofibromas, and schwannomas. Skeletal complications usually present early in life and can be attributed to abnormalities of bone growth, remodeling, and repair in NF-1 or can be secondary to nearby soft-tissue abnormalities complicating NF-1.

Skeletal complications can be categorized as generalized or focal manifestations [5]. Generalized skeletal abnormalities include osteoporosis/osteopenia, osteomalacia, shortness of stature, and macrocephaly. These features are common in individuals with NF-1, with decreased bone mineral density in both sexes reported in up to 50% of the patients, but usually mild [6–9]. Focal abnormalities of the skeleton are less common than generalized abnormalities, but may cause significant morbidity. Focal manifestations include spinal deformities, dysplasia of the tibia and other long bones, sphenoid wing dysplasia, chest wall deformities (pectus excavatum), dental abnormalities, periapical cemental dysplasia, and cystic osseous lesions. The effect of generalized abnormalities in the occurrence or progression of focal skeletal manifestations remains elusive.

The incidence of spinal deformities in association with NF-1 varies from 2 to 36% with scoliosis being the most common musculoskeletal manifestation of NF-1 [10, 11]. The purpose of this article is to present the spinal deformities that are most commonly associated with NF-1 and to identify the current management of spinal disorders based on the most recent literature.

2. Classification

Five distinct clinical forms of neurofibromatosis are currently accepted by most investigators: NF-1, NF-2, segmental NF, Legius syndrome, and schwannomatosis.

2.1 Neurofibromatosis type 1 (NF-1)

NF-1 or peripheral neurofibromatosis is a common autosomal dominant single-gene disorder with an estimated prevalence of 1:3,000 [12]. It is the most common form of neurofibromatosis and the one most likely to be encountered by the orthopedist. It is predicted to affect over two million people worldwide in all racial and ethnic groups. The NF - 1 gene is large in size, in the range of 350,000 base pairs with 59 exons, and its locus was discovered on chromosome 17q11.2 [12–14]. NF - 1 is a tumor-suppressor gene that encodes neurofibromin, a large cytoplasmic protein with 2,818 amino acids. Exons 21 through 27a encode a 360 amino-acid domain with homology with guanosine triphosphatase (GTP)-activating proteins (GAPs). The relevant domain, known as GAP-related domain (GRD), downregulates p21-Ras oncogene which promotes cell growth, proliferation, and differentiation. GAPs, including neurofibromin, inactivate Ras oncogene through their GTPase activity. Decreased synthesis or complete absence of neurofibromin expression, as in NF-1, results in unopposed activation of p21-Ras oncogene through GTP binding. This, in turns, leads to aberrant growth-promoting signals and the development of NF-1 associated neoplasms, including benign neurofibromas, malignant peripheral nerve sheath tumors, pheochromocytomas, and optic nerve gliomas, as well as to other clinical manifestations [15 ,16].

The NF - 1 gene displays almost complete penetrance. Individuals with NF-1 are constitutionally heterozygous for an NF - 1 gene loss-of-function mutation. Approximately 50% of affected individuals inherited the gene from an affected parent and 50% arise sporadically due to spontaneous mutations [16–19]. De novo mutations in the NF - 1 gene are associated with advanced paternal age [19].

The diagnosis of NF-1 is established when at least two of the most commonly presenting features of the disease as defined by the 1987 Consensus Development Conference of the National Institutes of Health are present (**Table 1**) [20]. In 97% of patients, a diagnosis is made by age 8 [21]. Molecular diagnosis with direct sequencing of the causative mutation is possible in 95% of patients with NF-1 and is indicated in uncertain cases and for prenatal diagnosis [22]. Differential diagnosis includes tuberous sclerosis and other conditions of pigmentation, such as McCune–Albright syndrome and mastocytosis. NF-1 is closely related to a number of other genetic syndromes

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TABLE 1. *Diagnostic criteria of NF-1*

1	Six or more café-au-lait macules more than 5 mm in greatest diameter in prepubertal individuals and more than 15 mm in postpubertal individuals
2	Two or more neurofibromas of any type or more than one plexiform neurofibroma
3	Freckling in the axillary or inguinal regions
4	Two or more Lisch nodules
5	Optic glioma
6	A distinctive osseous lesion, such as sphenoid dysplasia or thinning of long bone cortex, with or without pseudarthrosis
7	A first degree relative (parent, sibling, or offspring) with NF-1 by the above criteria

involving mutations of the Ras pathway, such as Noonan syndrome and LEOPARD syndrome.

2.2 Neurofibromatosis type 2 (NF-2)

NF-2 or central neurofibromatosis has an estimated incidence of 1 in 33,000 individuals and is associated with bilateral vestibular schwannomas and multiple spinal shwannomas [23, 24]. The NF-2 locus is located on the long arm of chromosome 22. Fifty percent of cases involve a new mutation. NF-2 is not associated with primary skeletal disorders; however, multiple paraspinal and intraspinal tumors (schwannomas and ependymomas) are common in this disorder. NF-1 and NF-2 are genetically distinct disorders with different gene loci, despite similarities in names.

2.3 Segmental Neurofibromatosis

Segmental neurofibromatosis is characterized by features of NF-1 involving a single body segment. Typically, only a single segment of the body (such as left upper extremity) is affected with café-au-lait spots and freckling, and lesions usually do not cross the body midline. Other segmental forms may involve deep neurofibromas in a single body segment.

2.4 Legius Syndrome

Early neurofibromatosis literature recognized that a mild form of NF-1 existed, consisting primarily of familial café-au-lait spots. In recent years, multiple families with such mild involvement have now been found to have mutations in the SPRED1 gene. Initially discovered by Legius et al. [25] this condition, now called Legius syndrome, can present with multiple café-au-lait spots, freckling, macrocephaly, and mild learning disabilities, but does not present with any of the benign or malignant tumors associated with NF-1. This condition is quite a bit less common than NF-1, with an estimated prevalence of about 1/50,000. Since patients with Legius syndrome can actually meet the clinical diagnostic criteria for NF-1, it can be appropriate to perform molecular testing if there is any question about diagnosis.

2.5 Schwannomatosis

Schwannomatosis is a distinct form of neurofibromatosis which typically involves multiple schwannomas throughout the body, but without the vestibular schwannomas typical of NF-2. Initially thought to represent a mosaic form of NF-2, it has now been determined that familial schwannomatosis is due to mutations in the INI1 gene, linked to NF - 2 on chromosome 22. It is a disease of adulthood that consists of multiple deep painful peripheral nerve sheath tumors that may occur in a generalized form or in a segmental distribution. Differential diagnosis from NF-2 can be difficult, and genetic testing of NF-2 and INI1 is now available to help in making this distinction.

3. Spinal Abnormalities in NF-1

3.1 Epidemiology

Spinal abnormalities are the most common orthope-

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TABLE 2. *Diagnostic criteria of dystrophic deformities*

1	Rib penciling
2	Posterior vertebral scalloping
3	Vertebral wedging
4	Spindling of transverse processes
5	Anterior vertebral scalloping
6	Widened interpedicular distance
7	Enlarged intervertebral foramina
8	Lateral vertebral foramina
9	Vertebral rotation
10	Paraspinal tumors
11	Dural ectasia

dic manifestation of NF-1. It is quoted as from 2 to 36% in the literature [10, 11]. In a report in 1988, Winter et al. [26] found only 102 patients having NF-1 by clinical criteria in a pool of approximately 10,000 patients with scoliosis. Functional scoliosis resulting from limb hypertrophy or long-bone dysplasia leading to limb length inequality must be ruled out in patients with NF-1. Rarely, unrecognized extrapleural thoracic tumors can present as focal scoliosis. These lesions are usually plexiform neurofibroma and are not visible on plain radiographs [27]. The spinal deformities tend to develop early in the life therefore, all preadolescent children with NF-1 should be evaluated by scoliosis screening or the Adam forward-bend test to rule out the presence of a spinal deformity.

It is important to emphasize that there is no standard pattern of spinal deformity in NF-1. All manner of spinal deformities in multiple planes and in any part of the spine may occur with NF-1 [28, 29]. The characteristic deformity tends to be a short-segmented, sharply angulated curvature that usually involves four to six vertebrae in the upper third of the thoracic spine [30]. We have traditionally classified the deformities into dystrophic or non-dystrophic types based on the coronal plane x-rays.

There are nine radiographic criteria most often used to classify the deformity as dystrophic. These include rib penciling (the rib being smaller in diameter than the second rib), vertebral rotation, posterior vertebral scalloping, vertebral wedging, spindling of the transverse process, anterior vertebral scalloping, widened interpedicular distance, enlarged interverteral foramina, and lateral vertebral scalloping. Recently, two more magnetic resonance imaging (MRI) findings have been added to the criteria used to classify the deformity as dystrophic: The presence of dural ectasia and the presence of paraspinal tumors (**Table 2**) [31]. More than three of these dystrophic features are considered diagnostic of dystrophic scoliosis. Nondystrophic curves are considered similar to idiopathic scoliosis.

3.2 Etiology

The cause of spinal deformity remains unknown. Several theories including metabolic bone deficiency, osteomalacia, endocrine disturbance, and mesodermal dysplasia have been proposed and are at best inconclusive [32-36]. The dystrophic changes may be attributed to intrinsic factors or may be associated with anomalies of the spinal canal secondary to abnormalities of the spinal cord dura mater.

Pressure erosive effects of dural ectasia and paravertebral tumors have been frequently found to be adjacent to and approximated to the deformities, initiating instability and subsequent deformity. Dural ectasia, a disorder unique to certain conditions, is an expansion or dilatation of the dural sac. The changes in the spinal canal induced by dural ectasia may increase the difficulty in obtaining adequate purchase for fixation of anchors during spinal deformity correction.

Scalloping was initially thought to represent the result of erosive pressure or direct infiltration of the vertebra by adjacent neurofibroma [37–41]. A neurofibroma-derived locally active biochemical substance or hormone that triggers dystrophic features in the adjacent vertebra has also been proposed [37]. The presence of an altered response of the vertebral bone in NF-1 to a paraspinal tumor has been hypothesized. An interactive pathophysiological mechanism between a genetically compromised bone and a neuroectodermal derivative, such as a contiguous

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neurofibroma or an abnormal meningeal sheath, is suggested by some authors [37, 39].

The etiological theory of vertebral scalloping being a primary developmental defect was supported by the presence of scalloping without adjacent lesions [42]. This was also supported by an MRI study in patients with NF-1, in which posterior vertebral scalloping was highly associated with dural ectasia, lateral scalloping was related to dural ectasia or neurofibromas in 50% of cases, and anterior scalloping was unrelated to dural ectasia or tumors [43]. The authors could not identify any association with dural ectasia or paraspinal tumors in more than one-third of their patients with MRI evidence of vertebral scalloping. Nevertheless, dural ectasia without associated vertebral scalloping was recorded in 10% of the cases.

A recent study in ten monozygotic twins with NF-1 demonstrated mixed concordance and discordance for presence of scoliosis [3]. The affected twin pairs were discordant for presence of dystrophic features, degree of curvature, and need for surgery. This finding suggests that both heritable and nonheritable factors contribute to the pathogenesis of spinal deformities in NF-1 patients. Dystrophic curves most likely require a nonhereditary event, such as an adjacent tumor or dural ectasia, or a second hit event in local bone cells leading to the underlying dysplasia. If occurrence and progression of dystrophic spinal deformity is affected by adjacent neurofi bromas, then therapies targeting to reduction or stabilization of paraspinal tumors could provide a promising approach to spine deformity prevention in patients with NF-1. Apart from its tumor suppressor activities through the Ras signaling, the role of neurofibromin pathways, such as bone morphogenetic protein (BMP) signal transduction [44]. This theory suggests that intrinsic bone pathology due to loss of a functional NF - 1 allele with subsequent Ras deregulation may be responsible for osseous manifestations in NF-1 through altered osteoblastic/osteoprogenitor differentiation, overgrowth of cellular tissue due to preferred fibroblast differentiation of mesenchymal cells, and impaired bony callus formation. Double inactivation of NF-1 by somatic mutation of the NF -1 gene in a population of cells which depends on neurofibromin-regulated Ras signaling to maintain normal bone was suggested to contribute to the occurrence or progression of tibia pseudarthrosis [45]. Although such second hit events have been demonstrated in pathological tissue from NF-1 tibias, it is unknown whether spinal deformities of NF-1 require a second hit event.

3.3 Imaging

Most often plain standing posterior-anterior and lateral radiographs are sufficient for screening the curvature. An angle of greater than 10° assigns the deformity as structural. When treatment is to be initiated, multiple planar films in supine bending modes and traction are necessary to determine flexibility. If there are adjacent structures requiring further clarification, higher levels of imaging are required, such as computed tomography (CT) for bony deformity or high- resolution contrast CT or MRI for soft tissue delineation.

3.4 Dural Ectasia

Dural ectasia is a circumferential dilatation of the dural sac which is filled with proteinaceous fluid. The slow expansion of the dura results in erosion of the surrounding osseous structures resulting in widening of the spinal canal, thinning of the laminae, and ultimately destabilization of the spine. Dural expansion through the neural foramina can cause meningoceles giving the radiographic dumbbell appearance. However, enlargement of a single neural foramen on an oblique radiograph is usually caused by neurofibroma exiting from the spinal canal rather than from the dural ectasia. Similar lesions are seen in other connective tissue disorders, e.g., Marfan's syndrome and Ehler–Danlos syndrome, although cause of these lesions in NF-1 is not known.

During this process, the neural elements are not affected. As a result of slow nature of this process and enormous widening of the spinal canal the neural elements have adequate room for accommodation, and there may be severe angular deformity and distortion without neurological deficit. The patients remain neurologically intact until later in the course of

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the disease process when destabilization of the vertebral column jeopardizes the neural elements. Dislocation of the vertebral column due to dural ectasia has been reported in the literature [46]. The destabilization at the costovertebral junction can result in penetration of the rib head into the spinal canal with neurological compromise [47, 48]. The presence of rib head or the neurofibroma in the spinal canal can result in intraoperative neurological deficit if instrumentation is used for correction of the curve without adequate decompression.

Dural ectasia can be readily seen on high volume CT myelography or contrast-enhanced MRI and is recommended before surgical intervention is undertaken for dystrophic curves. Higher imaging studies help to demonstrate extremely thin laminae. Surgical spinal stabilization and fusion does not alter the course of dural ectasia. Dural ectasia can result in failure of the primary fusion or the expanding dura ultimately can destroy a solid fusion leaving behind the instrumentation.

4. Cervical Spine Abnormalities

The cervical spine abnormalities in NF-1 have not received enough attention in the literature [49, 50]. Usually, the cervical lesion is asymptomatic. When the lesion is symptomatic, pain is the most common presenting symptom [51]. Cervical abnormalities are likely to be missed in presence of scoliosis or kyphoscoliosis of lower regions of the spine where the examiner's attention is focused on the more obvious deformity. In a study of 56 patients with NF-1, Yong-Hing et al. [52] reported that 17 patients (30%) had cervical spine abnormalities. Out of these, seven patients were asymptomatic, whereas the rest had limited motion or pain in the neck. Four patients had neurological deficits that were attributed to cervical instability. Four of the 17 patients required fusion of the cervical spine. Curtis et al. [53] described eight patients who had paraplegia and NF-1. Four of these patients had cervical spine instability or intraspinal pathology in the cervical spine.

The upper cervical spine should also be examined carefully. Isu et al. [54] described three patients with NF-1 who had C1–C2 dislocation with neurological deficit. All patients improved after decompression and fusion. We recommend that the cervical spine should be evaluated at the initial scoliosis assessment.

A lateral radiograph of the cervical spine is the initial screening tool. The NF-1 can be manifested on a plain radiograph in the form of dystrophic changes or malalignment [55]. If any suspicious area is noted on plain radiographs, right and left oblique views should be obtained to look for widening of the neuroforamina which may represent dumbbell lesions. MRI is the definitive study to evaluate these lesions.

Anteroposterior and lateral radiographs of the cervical spine should be obtained in all NF-1 patients who: (1) are placed in halo traction; (2) undergo surgery; (3) require endotracheal intubation; (4) present with neck tumors; (5) complain of neck pain; and (6) present with symptoms indicating intra- or extraspinal neurofibromas, such as torticollis or dysphagia [56]. If there is any suspicion of instability, CT and/or flexion- extension MRI are indicated. Erosive defects of the skull may be present in some patients with NF-1. Thus, plain radiographs of the skull prior to halo or Gardner–Wells tong traction pins application are strongly recommended.

The most common spinal abnormality in the cervical spine is a severe cervical kyphosis, which is often seen following a decompressive laminectomy without stabilization for an intraspinal lesion and is highly suggestive of the disorder [57]. We recommend stabilization of the spinal column at the same time of surgical removal of tumors from the spinal canal.

Ogilvie reported on the surgical treatment of cervical kyphosis by anterior fusion with iliac- crest or fibular bone graft or both [51]. He considered halo traction to be a useful preoperative step if the kyphosis was greater than 45°. In the presence of progressive cervical kyphosis, we recommend preoperative halo traction only if the deformity is flexible as judged by the radiographs. This should be followed by posterior fusion. If the deformity is rigid, then an anterior soft-tissue release followed by traction is safer.

Internal fixation with pedicle and lateral mass screws is preferred for posterior instrumentation. Sublaminar wire fixation may be difficult second-

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ary to dural ectasia and osseous fragility. For anterior fixation, we currently use bioabsorbable plates. Even with rigid instrumentation, postoperative halo immobilization is recommended until a fusion mass with trabecular pattern is seen on cervical CT.

5. Thoracic/Thoracolumbar Spinal Abnormalities

The two varieties of spinal deformity are well distinguished in these regions of the spine. Also, the natural history of spinal deformities is well studied for thoracic/thoracolumbar region. Patients more likely to develop progressive scoliosis of the thoracolumbar spine are children under 7 years of age who have thoracic lordosis (sagittal plane angle of less than 20° measured from T3 to T12) and paravertebral tumors. There is a strong association between modulation and progression of the spinal deformity. More specifically, curves that acquire either three or more penciled ribs or a combination of any three dystrophic features will almost certainly progress [28]. Other factors that have been associated with substantial curve progression include: 1) high Cobb angle at presentation; 2) early age of onset; 3) abnormal kyphosis; 4) vertebral scalloping; 5) severe apical rotation; 6) location of the apex in the middle-lower thoracic spine [34].

More recent MRI studies have questioned the theory of modulation [43]. Patients with radiographically labeled non-dystrophic curves have been found to have significant dysplastic changes on MRI. Having in mind the higher sensitivity of MRI in identification of dystrophic features than x-rays, we recommend characterization of the curve as dystrophic or not based on a combination of MRI and x-ray findings [31].

5.1 Non-dystrophic Scoliosis

This is the common variety of spinal deformity observed in NF-1. These curves behave similar to idiopathic curves with some differences [7, 9, 58]. This form usually involves 8-10 spinal segments. Most often, the deformity is convex to the right. However, these curves usually present earlier than the idiopathic curves and are more prone to progression. Furthermore, the rate of pseudoarthrosis following a fusion surgery is higher in these patients [49]. These differences can be attributed to the process of modulation and the underlying bone pathology. Compared to dystrophic curves, non-dystrophic curves tend to present in older children with less angulation and rotation of the deformity [59].

5.2 Dystrophic Scoliosis

This is an uncommon but malignant form of spinal deformity. It is characterized by early onset, rapid progression and is more difficult to treat [60, 61]. Typically, the dystrophic curve is a short-segmented, sharply angulated type that includes fewer than six spinal segments. Dystrophic curves may be associated with kyphosis and have a higher incidence of neurological injury [61, 62].

Dystrophic vertebral changes develop over time (**Table 2**).

5.2.1 Natural History

The onset of spinal deformities may occur early in patients with NF-1. Usually early onset scoliosis is associated with kyphosis giving rise to kyphoscoliotic deformities. Calvert et al. [63] presented a series of treated ($n = 34$) and untreated ($n = 32$) patients who had NF-1 and scoliosis. Seventy-five percent of patients in the nontreated group had kyphoscoliosis. The investigators reported that patients, who had severe anterior vertebral scalloping noted on the lateral view, progressed an average of 23° per year for scoliosis and kyphosis.

Some of the non-dystrophic curves exhibit the phenomenon of modulation. Durrani et al. [28] defined modulation as a process by which a nondystrophic curve acquires the features of a dystrophic curve and behaves as a dystrophic curve. They reported that modulation occurred in about 65% of their patients. Modulation occurred in 81% of patients who presented with scoliosis before 7 years of age and in 25% of those diagnosed after 7 years of age. Rib penciling acquired through the modulation period was the only factor that was statistically significant in influencing the progression of the deformity. These authors based their report on plain radiographic findings. Some of the recent reports with the use of MRI of spine have shown the presence of dystrophic findings in the spine before they are apparent on

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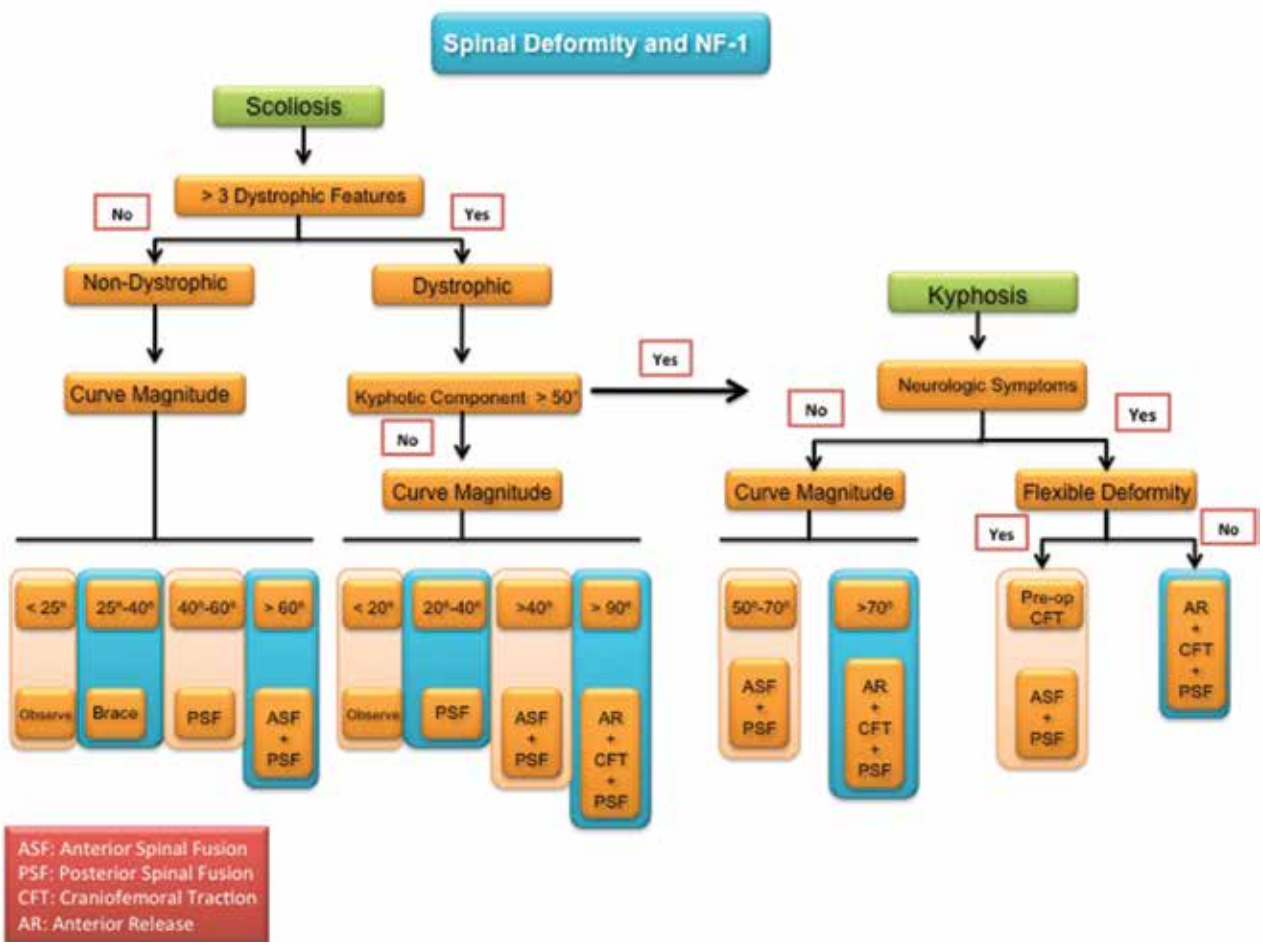

Fig. 1. *Spinal deformity and NF-1*

the plain radiographs. Based on these reports, it can be speculated that true modulation may be rare, and many of the apparent non-dystrophic curves are actually dystrophic curves which subsequently present themselves with radiographical changes of dystrophic curve giving an impression of modulation.

A retrospective review of 694 patients with NF-1 revealing 131 patients (19%) with a scoliosis ranging from 10° to 120° was performed [31]. Mean age at diagnosis of scoliosis was 9.0 years, with 18 patients (15%) having onset before 6 years of age. Forty-six patients (35%) required surgical repair, usually anterior and posterior spinal fusion with instrumentation. Six patients had growing rods successfully placed. Tumors near the spine were found in 65 % of patients requiring surgery.

It is well known that despite apparent solid fusion, some dystrophic curve shows progression. This tendency is more noted in patients with kyphosis (>50°). The vertebral subluxation, disk wedging, and dystrophy of peripheral skeleton are other factors associated with progression of the deformity after fusion [64].

5.2.2 Treatment

The treatment of non-dystrophic curvatures is very similar to idiopathic scoliosis. The curve of less than 25° should be observed (**Fig. 1**). Curves between 25° and 40° can be treated with brace successfully [35]. Once beyond 40°, surgery by posterior spinal fusion is usually indicated [65]. Curves >55°-60° are treated with anterior release with bone-grafting, followed by an instrumented posterior spinal fusion [49]. This is necessary because the curve is usually more rigid than is a similar-sized curve in idiopathic scoliosis. We recommend postoperative orthotic immobiliza-

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tion, although others have managed these patients without postoperative immobilization, with good early results [29].

Dystrophic curvatures of less than 20° should be treated by observation. Serial spinal radiographs at 6-month intervals should be obtained to check for progression of the deformity [49]. Bracing of progressive dystrophic curvatures is ineffective and surgery is usually recommended [10, 35, 66]. For adolescent patients with dystrophic curvature greater than 20°–40° of angulation, a posterior spinal fusion with segmental spinal instrumentation is recommended [10, 62]. In more severe dystrophic scoliosis, anterior fusion should be performed in addition to posterior fusion, to increase the fusion rate, and to reduce the risk for progression despite solid posterior fusion. Preoperative halo traction may be beneficial for the treatment of severe curves, including those with kyphoscoliosis [10, 58, 67, 68]. It allows gradual and controlled soft tissue relaxation and curve correction before surgery or between staged surgeries; however, it is contraindicated in patients who have cervical kyphosis. Daily neurological evaluations are mandatory to avoid spinal or cranial nerve injuries. Nutrition is also paramount during this time. We use supplemental nasojejunal feeding in between stages to decrease the protein depletion that is seen in staged patients [34, 69]. We recommend anterior release, nasojejunal tube alimentation, and craniofemoral traction for rigid curves of >90°.

The dystrophic curves that are present in late juvenile and early adolescent period pose a challenge to the surgeon. These curves have a high rate of pseudoarthrosis following a posterior spinal fusion [49, 61, 65]. A combined anterior and posterior spinal fusion has been recommended in these patients to decrease the rate of pseudoarthrosis and crank-shaft [70-73]. In our experience, an early fusion of the spine in this age group does not significantly alter the final height and its benefit outweighs the risk of severe progression. It is suggested that the primary reason for fusion failure is an inadequate anterior procedure [74]. However, erosion from enlarging neurofibromas, dural ectasia, and meningoceles may play a role.

Dystrophic curves in infants, toddlers, and early juvenile patients present even more of a challenge. In this age group, a spinal fusion can certainly have a significant effect on overall height as well as the size of the thoracic cage. Smaller size of the vertebrae can pose difficulty in the instrumentation. On the other hand, progression of the curve itself can significantly distort the thoracic cage which can lead to cardio-thoracic decompensation. Most centers recommend observation initially for spinal deformities to determine whether or not it will progress. If the child is very young (under 5–6 years), a corrective cast or bracing may be attempted, most often with little to marginal success. However, it may allow the surgeon to buy some time. Growing rods have been used to obtain correction without definitive fusion and to lengthen or "grow the spine" every 6 months, but with varying success and a high rate of complications.

5.2.3 Growing Rod Instrumentation

The growing rods have been used successfully in the treatment of early onset idiopathic curves.

These devices have been shown to prevent the progression of the curve while preserving the longitudinal growth of the spine [75]. The currently available dual growing rods have been shown to be superior to the previous versions of submuscular single growing rods [76]. We have used dual growing rods on early onset dystrophic curves with a great deal of optimism [62].

The routine lengthening is made at 6-month interval. The use of growing rod instrumentation in NF-1 is also associated with a high incidence of complications. The high rate of complications has also been reported for idiopathic patients (75). The most common complication we have encountered is proximal junctional kyphosis. This is especially common in the patients with high thoracic or cervicothoracic curves. Other complications encountered are infection and rod breakage.

Although the use of growing rod instrumentation is associated with higher complication rate, its benefits outweighs the risk in patients with early onset dystrophic scoliosis. Our early results with the use of growing rods remain encouraging. This is a promising technique made especially useful because most dystrophic curves have early onset.

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6. Other Spinal Deformities

6.1 Kyphosis

Kyphoscoliosis is defined as scoliosis accompanied by a kyphosis of greater than 50°. It may occur by gradual scoliotic rotation and progression or it can be found early in the disease with an abrupt angular kyphotic curve [78]. Vertebral bodies may be deformed so severely that they are confused with congenital deformities. Severe kyphosis is the most common cause of neurological deficits in NF-1 [62]. Use of traction in patients with rigid and severe kyphosis can increase the tension on the spinal cord leading to neurological deficits. Traction following anterior release is safe when monitored appropriately. For curves greater than 50°, anterior surgery (release and fusion) is recommended, followed by posterior segmental instrumentation one or two levels above and below the end vertebrae [32, 49, 58, 64]. Assessment of the fusion mass by CT at 6 months postoperatively is recommended. If pseudarthrosis is noted, augmentation of the fusion mass is indicated.

We recommend that the anterior procedure should be undertaken from the convex side of the deformity, since the exposure is extremely difficult from the concave side [79]. The anterior fusion should include the entire structural area of the deformity with complete disk excision and local strut grafting. Multiple grafts or cages should be placed into the vertical weight-bearing axis of the torso, with the strong autologous fibula or rib graft placed more anteriorly [62, 66]. Strut grafts should have contact with each other and with the vertebral body to prevent resorption noted when graft material is surrounded by pathological tissue. Anterior release and fusion should be followed by posterior instrumented fusion using a large amount of autologous iliac crest bone graft and BMP in selected cases.

6.2 Lordoscoliosis

Lordoscoliosis has not been so frequently reported in patients with NF-1 compared to kyphoscoliosis. However, lordosis of the thoracic spine predisposes to significant respiratory compromise and mitral valve prolapse [77, 80].

Anterior release and intervertebral fusion followed by posterior instrumented fusion is considered as the most reliable surgical option to achieve correction of dystrophic lordoscoliosis [32].

6.3 Spondylolisthesis

Spondylolisthesis in patients with NF-1 is rare. It is characterized by pathological forward progression of the anterior elements of the spinal column. Spondylolisthesis in patients with NF-1 is most often associated with pathological elongation and thinning of the pedicles or pars interarticularis by lumbosacral foraminal neurofibromas or dural ectasia with meningoceles [32]. The vertebral bodies may also be small and dystrophic.

Fusion may also be delayed because of the forward traction effect of the vertebral bodies and the slow healing and remodeling of bone in NF-1. We recommend a combined anterior and posterior fusion from L4-to-sacrum using intervertebral body grafting and lumbosacral instrumentation. Postoperative immobilization is indicated until the fusion is absolutely solid.

7. Conclusion

NF-1 is the most common human single-gene disorder. Skeletal complications usually present early in life and can be attributed to abnormalities of bone growth, remodeling, and repair in NF-1 or can be secondary to nearby soft-tissue abnormalities associated with NF-1. Scoliosis is the most common osseous manifestation of NF-1. It is important to recognize the dystrophic curve and to distinguish it from the non-dystrophic curve.

The management of spinal disorders in young children in NF-1 continues to be problematic. The use of growing rods allows more longitudinal growth than fusion and more life freedom than bracing. The problems we have encountered are mechanical and could be expected when proximal and distal fixation is performed over an otherwise completely mobile spinal column. The multiple surgeries increase the potential for complications including infections. We continue to pursue solutions to our problems.

Conflict of interest:

The authors declared no conflicts of interest.

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ΠΕΡΙΛΗΨΗ

Η νευροϊνωμάτωση τύπου 1 (NF-1) αποτελεί την πιο συχνή μονογονιδιακή ανωμαλία στον άνθρωπο. Οι σκελετικές επιπλοκές συνήθως παρουσιάζονται νωρίς και μπορούν να αποδοθούν στις ανωμαλίες της οστικής ανάπτυξης, ανακατασκευής και επιδιόρθωσης στην NF-1 ή μπορεί να είναι απότοκες των ανωμαλιών των γειτονικών μαλακών μορίων που σχετίζονται με την NF-1. Η σκολίωση αποτελεί την πιο συχνή σκελετική εκδήλωση της NF-1. Είναι σημαντικό να αναγνωριστούν οι δυστροφικές παραμορφώσεις και να διαχωριστούν από τις μη δυστροφικές. Η διαχείριση των σπονδυλικών παραμορφώσεων στα μικρότερα παιδιά με NF-1 παραμένει προβληματική. Η χρήση των εκπτυσσόμενων ράβδων επιτρέπει την κεφαλουραία ανάπτυξη της σπονδυλικής στήλης και παρέχει μεγαλύτερη ελευθερία συγκριτικά με τον κηδεμόνα. Τα προβλήματα είναι κυρίως μηχανικής φύσεως και παρατηρούνται όταν πραγματοποιείται κεφαλική και ουραία σταθεροποίηση σε μια τελείως ασταθή σπονδυλική στήλη. Οι πολλαπλές επεμβάσεις αυξάνουν την πιθανότητα επιπλοκών όπως οι λοιμώξεις. Σκοπός αυτού του άρθρου είναι να παρουσιάσει τις σπονδυλικές παραμορφώσεις που σχετίζονται με την NF-1 και να αναλύσει την αντιμετώπισή τους έχοντας ως βάση την πιο πρόσφατη βιβλιογραφία.

ΛΕΞΕΙΣ ΚΛΕΙΔΙΑ: νευροϊνωμάτωση, σκολίωση, κύφωση, δυστροφική παραμόρφωση, NF-1

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Hip injuries in skeletally immature athletes

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ABSTRACT

Nowadays, children play no more in the outdoors, while they regularly participate in competitive activities, being guided by specialists. As compared with the past, injuries are bigger in number, since several tissues are more often being exposed to external risk and obviously they are more intensely overworked. The area of pelvis and hip is more often affected. Transient synovitis is the most common cause of painful limpness to the athlete child, apart from other potential severe situations (such us slipped capital femoral epiphysis, Legg-Calvé-Perthes disease, the avulsion fractures or stress fractures of femoral neck) which require our attention. Prevention has reduced the incidence of these injuries and it is considered more important than any other form of therapy. Parenthood is also very important in order to manage the situation. Doctor's advice should be realistic and full of altruism and affection, because children's delicate Psyche should be always respected.

KEY WORDS: sport injuries; hip; pelvis; child

1. Introduction

All scientists claim that moderate natural activity on a systematic basis and in a normal way, has a great amount of advantages for children. Although, international bibliography does not manage to support the benefits to the musculoskeletal system, does not hesitate to admit that natural activity provide positive results with regard to the arterial pressure, the control of obesity and the decrease of serum lipids [1-5]. Regarding to the adults, natural activity combines with low incidence of cardiovascular disease, diabetes type II, osteoporosis, colon cancer and breast cancer.

Doctor's role is very essential. He is called on recommending natural activity to his patients and make all the necessary adjustments to the type of exercise

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according to their condition of health. A systematic training program should be a way of life, especially for those people with low natural activity and reduced sports participation, such as deficient children in social skills and those having a low social-economic status [6]. Apart from suggesting the participation in sports, doctor should also be responsible to choose young athletes according to scientific tests, in addition to the diagnosis and therapy of various injuries [7].

2. Epidemiology and prevention of injuries

In the USA, with a population of 320 million people, almost 30 million children and teenagers take part in competitive activities. Nearly three million injuries are responsible for abstaining from sports [8,9]. Generally, the incidence and the gravity of injuries increase by the age and during adolescence, while they are also related to speed, power and intensity of competition. The acknowledgement of mechanisms that provoke sports injuries, in addition to the promotion of norms which can reduce the possibilities of these mechanisms (such as the punishment of dangerous game) have reduced the frequency of serious trauma. It has also been reduced by removing environmental risks, like trampoline in gymnastics [10].

An injury, sometimes, may be relapsed because of its incomplete treatment. Full recovery reduces the incidence of injuries. In young athletes, a systematic training before the competing period, especially when a great importance is given to spread, flexibility, jumping, springiness, is accompanied by an impressive reduction of the incidence of injuries in sports extremely dangerous, such as football. It has been proven that traditional stretching or massage do not reduce the risk of injury or the feeling of muscle fatigue [11-14]. Preventive physical exam of the child before his participation in various sports, is a good supply in order to define the preventive strategy that, in any case, should be followed [15].

3. Sports injuries treatment

3.1 Acute injuries

Most musculoskeletal injuries are related with strains, ligament failure and muscle strains [7]. The history is not always clear. Serious injuries which usually suggest the presence of a structural disorder, give symptoms and clinical findings, such as swelling, deformation, weakness for the continuation of sports, limpness, painful snapping, mechanical joint locking, or the sense of instability. Some concepts should be clear: The *strain* represents the injury of a ligament or an articular capsule, the "*twitch*" has to do with a muscle or a tendon, the *muscle strain* is referred to a crushed injury of any soft tissue.

Strain is classified in grades 1-3. Grade 1 means that some fibers have been ruptured, although during physical exam no instability of the ligament is found. In grade 2, most fibers have been ruptured, resulting in some instability of the ligament. Grade 3 implies that all fibers have been ruptured and during clinical exam total instability of the ligament is perceptible. The *muscle strain* is also classified in grades 1-3. Grade 1 causes moderate soreness during the exam of the muscle, in addition with minimal muscle weakness. Grade 2 causes a higher intensity pain and moderate weakness during the muscle exam. Grade 3 means total muscle or tendon tear and causes pronounced weakness and palpable gap on the muscle belly or on the tendon itself.

3.2 Overuse injuries

These injuries are caused by repeated microtrauma which exceed healing potential of the tissues [7]. They concern muscles, tendons, bones, capsules, cartilages and nerves. Those injuries may by observe in all sports, but they are more frequent in sports which require repetitive movements (such as running, swimming, tennis, gymnastics).

Causal factors are distinguished in extrinsic (trainer mistakes, lacking equipment, unsuitable exercise surface) and intrinsic (anatomical structure of the athlete, and/or pathological processes) [16]. Mistakes during sports activity are most perceptible predisposition factors. As usual, at the beginning of the competitive period, athletes break the "rule of 10%" (*it is not allowed to increase the time or intensity of training more than 10% weekly*).

Intrinsic factors are related with biomechanical disorders because of leg length discrepancy (LLD), pes

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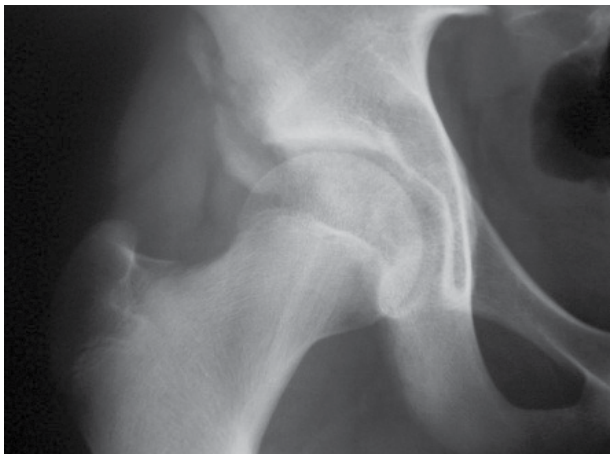

Fig 1. *In a 15-years-old boy with acute pain on right groin during running, the radiography showed an avulsion fracture of anterior superior iliac spine and the acetabular ridge due to intense pull of rectus femoris muscle*

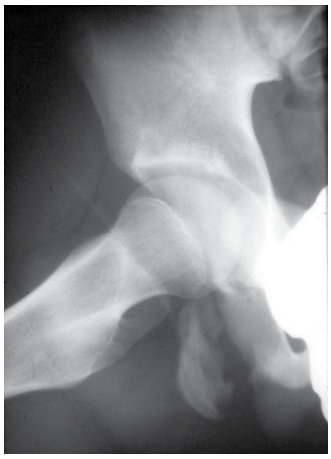

Fig 2. *Avulsion of ischial tuberosity due to intense pull of hamstrings in a 13-years-old sprinter. The diagnosis was carried out with only the radiological assessment, in accordance with the information from history and clinical examination*

planus, cavovarus foot, tarsal coalition, calcaneovalgus foot, external tibial torsion, or femoral anteversion. They also are related with muscle imbalance, inflexibility of muscles, as well as with other various pathological situations (such as lacking physical condition, inadequate nutrition, amenorrhea, obesity) [7,17].

During medical interview, the little athlete gives information about the special characteristics of the sport. For example, runners should be asked about the shoes, the orthotics, the surface on which they run, the number of kilometers which they run every week (or the time they spend practicing every week), the intervals trainings, or the training who made on the mountain, in addition with previous injuries and periods of rehabilitation [18]. When all factors are appreciated, there is a break (or a modification) of the sport in order to avoid the recrudescence of the injury.

For hard–training athletes who develop an overuse injury, it is not considered necessary to stop exercising. As a rule, it is advised a program of rehabilitation which allows the athletes to return to sports as soon as possible, and at the same time they are not vulnerable in front of a possible relapse [19]. Early appreciation of an overuse injury requires changes to a smaller degree in training program.

One of the goals of the treatment is to control the pain and the muscle soreness, and secondly flexibility, strength, endurance and proprioception must be restored [20]. In many injuries caused by overuse, the inflammation has an insignificant role. For example, in most tendon injuries the term "*tendonitis*" is considered obsolete, since the pathology of the tendons does not reveal the presence of inflammation. On the contrary there is a scar tissue devoid of vessels. Most of these entities are now called "*tendinosis*", and the administration of anti-inflammatory drugs in them is considered insignificant, except for their analgesic effect.

3.3 Injuries of pelvis and hip

Hip and pelvic injuries represent only a little amount of sports injuries, but they are often severe and they require appropriate and early diagnosis [19]. Hip diseases are often manifested by knee-ache, without pathological signs from the physical exam of the knee.

In children who do physical activity, the ***transient synovitis*** is the most common cause of pathology of the hip. It is usually manifested by acute onset pain and limpness [21,22]. The child refuses to use the suffering extremity, while during physical exam he

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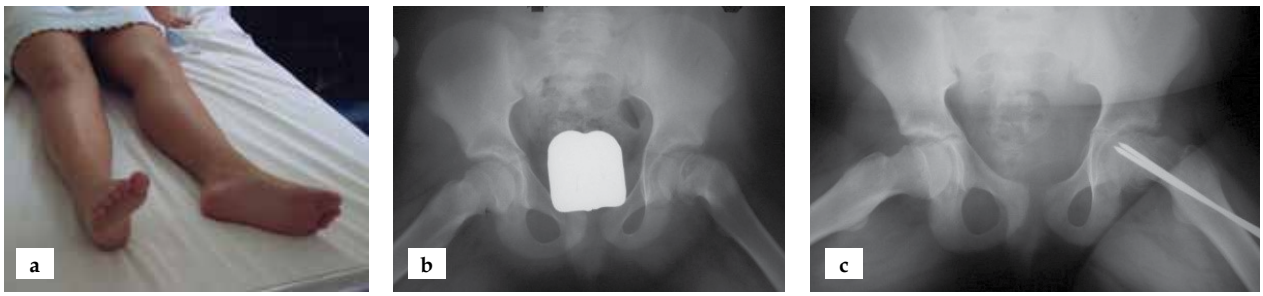

Fig. 3. *Slipped left capital femoral epiphysis* ***a.*** *Clinical signs* ***b.*** *Preoperative radiography and* ***c.*** *Postoperative radiography showing the management of epiphysiolisthesis in situ*

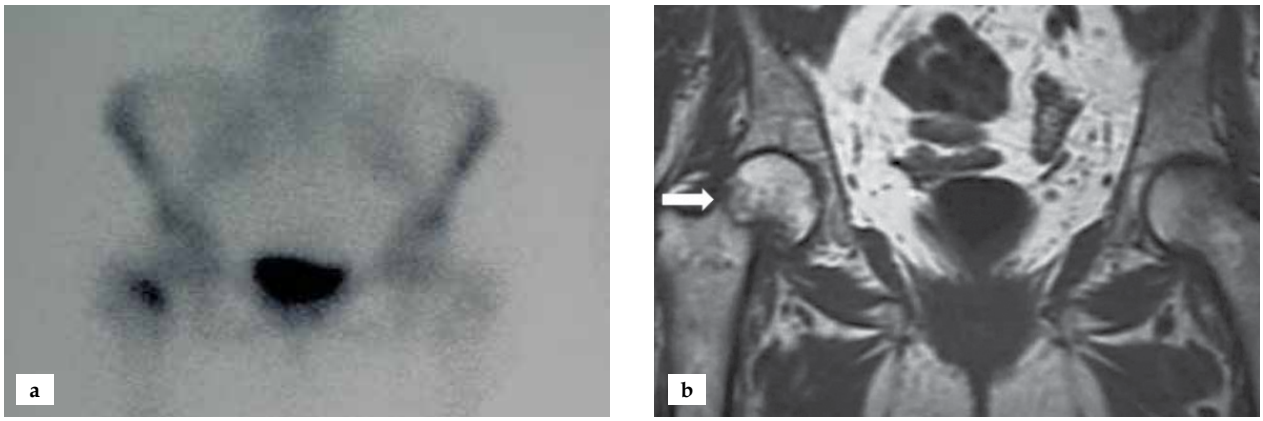

Fig. 4. *Stress fracture of right femoral neck of a young athlete with no obvious findings* ***a.*** *An increased concentration of radioisotope during bone scanning* ***b.*** *MRI shows the fracture (arrow)*

seems to suffer when he moves the hip. In the history, a minor injury is being reported. It is about a self-limiting process, usually restored between 48-72 hours after onset.

The ***Legg-Calvé-Perthes disease*** is also appeared during childhood, with gradual onset of limpness and hip ache [23]. Predisposing factors of the disease are hyperactivity or lack of concentration of the child, in addition with various nutritional habits.

Till the skeletal maturity, young athletes are vulnerable to injuries of apophysis (for example, anterior superior iliac spine). The ***iliac apophysitis*** is the most common in this area and it appears whether as a result of overuse, or by direct blow [22,24]. Treatment is, basically, conservative and it requires the temporary restriction of activities.

The ***avulsion fractures*** are mostly related to teenagers who participate in sports requiring a sudden beginning (**Fig. 1**). As soon as large muscular groups are contracted, the applying forces are greater than those which bones can tolerate [25,26]. Frequent sites of avulsion fractures are the anterior superior iliac spine (due to traction of sartorius), the anterior inferior iliac spine (rectus femoris), the lesser trochanter (iliopsoas) and the ischial tuberosity (posterior femoral muscles). The symptoms are located pain, tenderness, and swelling, loss of muscular strength and limitation of hip motion. Radiograms are necessary (**Fig. 2**). The initial treatment includes ice, analgesics, rest and exercises without causing pain. It is usually recommended the use of crutches for patient's ambulation. Surgical treatment is rarely required, since most of these fractures are satisfactorily healed, even if there is an important displacement.

The ***slipped capital femoral epiphysis*** often appears in ages 11-15 years old, during growth spurt in

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the adolescence (**Fig. 3**). Apart from traumatic causes, hormonal factors are also implicated [16]. Treatment must be operative. Osteonecrosis of the femoral head is the most frequent complication [27,28].

Stress fractures of the femoral neck are usually manifested by progressively increasing pain in the area of hip, in children who practice resistance sports[29]. Girls are more vulnerable, especially those who manifest the so-called "Athletic Triade", meaning the combination: alimentary disorders-secondary amenorrhea-secondary osteoporosis. During physical exam, pain is present every time that flexor hip muscles are contracted or torsional movements of the hip are realized. If x-rays do not succeed in evidencing the characteristic periosteal reaction (compatible with the stress fracture), then bone scan and MRI are required (**Fig. 4**). It is recommended to visit an Orthopaedic Surgeon, because stress fractures of femoral neck tend to conduce on nonunion or displacement, due to an insignificant trauma or continued bearing, since diagnosis is late [30,31]. Displacement may, more often, occurs to the cortex which supports tensile forces and, more rarely, to the opposite cortex which get compressive forces. This fact justifies preventive internal fixation of the fractures of the first group. These fractures are implicated for high risk of osteonecrosis of the femoral head [32,33].

The ***osteitis pubis*** represents an inflammation of pubic symphysis and it is caused by repetitive trauma during the broadside movements of pelvis. It appears in every athlete who participates in running activities, but it is more frequent to sports which require an action of the adductors muscles of hip, like hockey and football[34]. Athletes usually feel a vague, ill-defined pain in anterior pelvic region, unilateral or bilateral. On physical exam, a tenderness directly over the pubic symphysis, and sometimes to the central part of adductors muscles, is palpated. Any test which requires adductors contraction, evokes pain. There are no specific radiographic features (areas of bony erosions or sclerosis, diastasis of pubic symphysis with osteolysis), excepting the symptoms insist for 6-8 weeks. Bone scan shows an increased activity in area of pubic symphysis, while MRI can demonstrate an early bone marrow edema. Rest for a period of 6-12 weeks will be useful. Some patients turn to steroid injections, as a supplementary treatment.

Tears of the acetabular labrum may occur to the hip at the same way as the corresponding tears of the glenoid labrum happen [35]. The patient may refer a previous trauma, while he complains about an acute pain to the anterior surface of the hip, associated with a sensation of snapping or locking. Physical exam is not sufficient for the diagnosis. MRI arthrogram is the study of choice and has a sensitivity of 92% for detecting labral tears [22].

The ***snapping hip*** is caused whether by iliopsoas tendon sliding over anterior bursa of the hip, or by the iliotibial tract sliding over greater trochanter. It happens frequently to ballet dancers and runners in their teens. It may happen as a result of overuse (more frequent), or as an acute injury [36]. Athletes have a painful or painless snapping sensation located to the external or/and anterior surface, deep inside the articulation. Physical exam often reproduce the symptom. X-rays are not helpful. Treatment includes analgesics, rest, bio-mechanical assessment, and stretching and flexibility exercises [37]. The athlete returns to his sport activities as soon as possible.

Hip bursitis is caused by inflammation of any bursa which surrounds the hip, such as the insertion of iliopsoas on the lesser trochanter, the insertion of middle gluteal muscle on the greater trochanter and the origin of knee's flexors muscles on the ischial tuberosity [38]. By rule, treatment is conservative and it includes exercises following a specific weekly algorithm (stretching, isometric, isotonic with or without bearing).

4. Useful recommendations to parents

Children should not be forced to participate in a sport, just because ergonomic physiological control revealed that they can have a satisfying efficiency [7,16]. On the other side, they should not be excluded by their favourite sports just because their constitutional issues showed that they might be physically impaired. Unreasonable press by the parents and coach define if the child will be happy with this sport, and if he will continue to practise it during and after adolescence.

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Parents who give great importance to victory bring stress to their child until he arrives to believe that he does not deserve it. He prefers not to participate in sports in order to avoid the delusion of failure. Children should learn how to lose and we should teach them the way to get over the feeling of failure, assuming to help them trying again.

Coach and sports teacher are essential. If they have their own lack of self-confidence, then it is obvious that they will keep on trying the athlete child beyond his personal limits, aiming only for winning, just because victory means recognition. Children do not need to be surrounded by selfish people. Only if they socialize with mature people they will have the opportunity to maturate normally.

Conflict of interest:

The authors declared no conflicts of interest.

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ΠΕΡΙΛΗΨΗ

Τα παιδιά στις μέρες μας έπαψαν πλέον να παίζουν ελεύθερα σε ανοικτές αλάνες, ενώ συμμετέχουν συστηματικά σε οργανωμένα αθλήματα υπό την καθοδήγηση εξειδικευμένου προπονητή. Σε σύγκριση με το παρελθόν οι τραυματισμοί είναι περισσότεροι, καθώς οι διάφοροι ιστοί εκτίθενται συχνότερα σε εξωτερικούς κινδύνους και σαφώς καταπονούνται εντονότερα. Η περιοχή του ισχίου και της λεκάνης προσβάλλεται συχνά. Η παροδική υμενίτιδα είναι η συνηθέστερη αιτία επώδυνης χωλότητας στο αθλούμενο παιδί, πλην όμως διάφορες άλλες δυνητικά σοβαρότερες καταστάσεις, όπως η ολίσθηση της άνω μηριαίας επίφυσης, η νόσος των Legg-Calvé-Perthes, τα αποσπαστικά κατάγματα ή τα κατάγματα κόπωσης του μηριαίου αυχένα, απαιτούν την προσοχή μας. Η πρόληψη έχει μειώσει τη συχνότητα αυτών των κακώσεων και θεωρείται σημαντικότερη από οποιαδήποτε μορφή θεραπείας. Στην αντιμετώπιση επίσης σημαντικό ρόλο μπορούν να παίξουν οι ίδιοι οι γονείς. Οι συμβουλές του ιατρού οφείλουν να στηρίζονται σε ρεαλιστική βάση και να διαπνέονται από αίσθημα αλτρουισμού και αγάπης, καθώς ο ευαίσθητος ψυχισμός των παιδιών πρέπει πάντα να γίνεται σεβαστός.

ΛΕΞΕΙΣ ΚΛΕΙΔΙΑ: αθλητικές κακώσεις, ισχίο, λεκάνη, παιδί

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CLINICAL CASE

Ultrasonography: An alternative imaging modality in diagnosing greenstick fractures. Early experience in a county hospital

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ABSTRACT

Aim: In this case series clinical protocol, comparison of X-ray examination vs ultrasonography in detecting minimally displaced distal radius fractures in children is presented.
Patients and Methods: In twenty four children, 2 to 14- year old, with a suspected fracture of the distal radius ultrasound of the wrist was applied prior to X-ray. Tenderness on palpation and the point of maximal tenderness was located and marked. This was the place where the probe was first placed. The bone was examined circumferentially. In cases where the children could not cooperate, the examination began at the wrist area and the probe was moved proximally.
Results: In all greenstick fractures pain on palpation was present. Whenever there was a fracture diagnosis with the X-rays, the ultrasound was also positive for fracture.
Conclusion: From the data of the above study, it seems that ultrasound, is at least as sensitive as radiography in detecting greenstick fractures. Therefore it is an alternative, low-cost, safe, effective and sensitive bedside test in order to reveal minimally displaced fractures in children.

KEY WORDS: torus fractures; greenstick fractures; distal radius fracture; ultrasonography

1. Introduction

Children have a thick periosteum that protects the bony cortex which is softer than in adults. This is the reason that some types of fractures such as greenstick, buckle or torus, long bone bowing, are seen only in children. A torus fracture is a bulging of the bony cortex and happens when compressive forces act on the bony metaphysis. In a greenstick fracture there is a clear break of the convex surface and bending of the concave one and happens by more severe forces [1]. Since the distinction between these two types of fractures might be confusing, and the treatment is more or less the same, from

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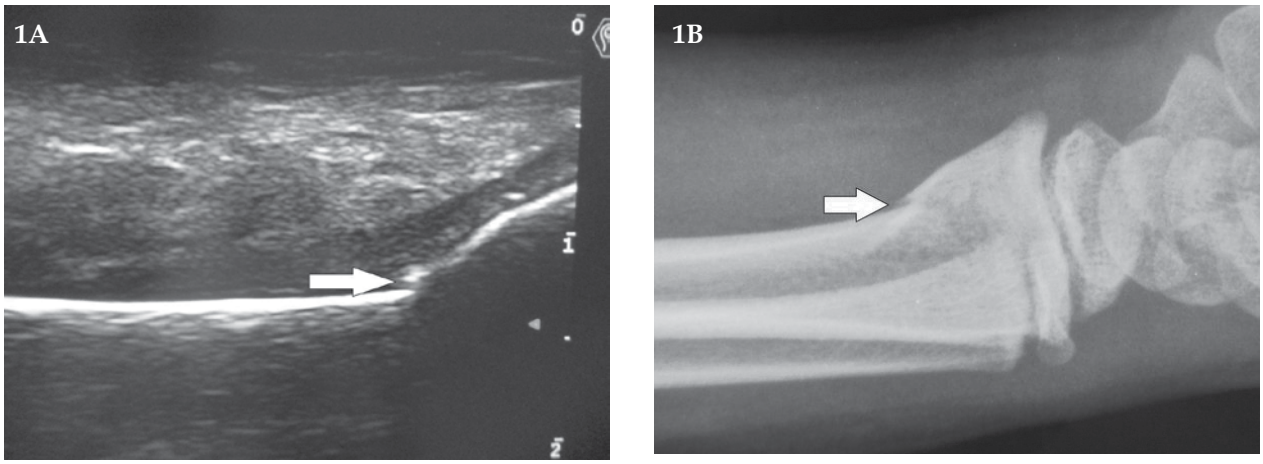

Fig. 1. *The arrow in both figures (ultrasound 1a and x-rays 1b), shows a green stick fracture*

now on they will be referred as minimally displaced distal radial fractures. The diagnosis of minimally displaced fractures of the distal radius in children can be established by history, physical examination and plain X-rays. The latter is still the gold standard amongst the imaging modalities to detect these fractures. However, there are concerns regarding radiation exposure and an alternative imaging tool for the diagnosis of these common fractures in children remains unsolved. Musculoskeletal ultrasonography was first used in rheumatic diseases in the 80's. Since then it has been used in several specialties including Orthopedics. Musculoskeletal ultrasound has been recently used in Orthopedics and there are still a lot to be done in that area [2].

This study aimed to investigate if the ultrasound can add to the diagnosis of minimally displaced fracture of distal radius or even replace the gold standard of plain X-rays investigation.

2. Patients and Methods

Children up to14 years old complaining of wrist pain after a fall were included. On physical examination there was tenderness on palpation over the distal radius. However, no obvious deformity of the wrist or forearm was noticed.

All patients had an ultrasound scan of the injured limb at the time of presentation in A&E using an Esaote My lab 70 X-vision and a linear probe (13 MHz). The bone was examined circumferentially, to avoid false negative results. After the ultrasound investigation, they had an X-ray of the wrist area (postero-anterior and lateral views) and the films were interpreted by a radiologist and compared to ultrasonography results. The X-rays diagnosis was performed by a specialist radiologist and it was then compared to the ultrasound diagnosis made by the authors.

3. Results

Twenty four patients presented in A&E Department met the inclusion criteria. There were ten females and fourteen males with a mean age of 8 years (2 to 14). The presentation time after the injury ranged from hours to three days. The point of maximal tenderness on palpation was marked. This was the site where the probe was first applied. The bone was examined circumferentially. In cases where the child could not cooperate, the examination started at the wrist area and the probe moved proximally.

The parents were informed about the study and a verbal consent was obtained. All parents, consent to have their kids an ultrasound examination and all children relaxed quite soon after the application of the probe onto their wrist when realized it was not painful.

The area where the fractures occurred is shown in **figure 1** (arrow) In some cases the fracture side was also examined using Doppler trying to allocate the epiphyseal nutrient vessel (**Fig. 2**) All fractures were within the criteria set for the minimally displaced distal radial fracture (**Fig. 2 and Fig. 3**).

In all cases there was a fracture diagnosis with the use of X-ray, the ultrasound was also positive for such a fracture.

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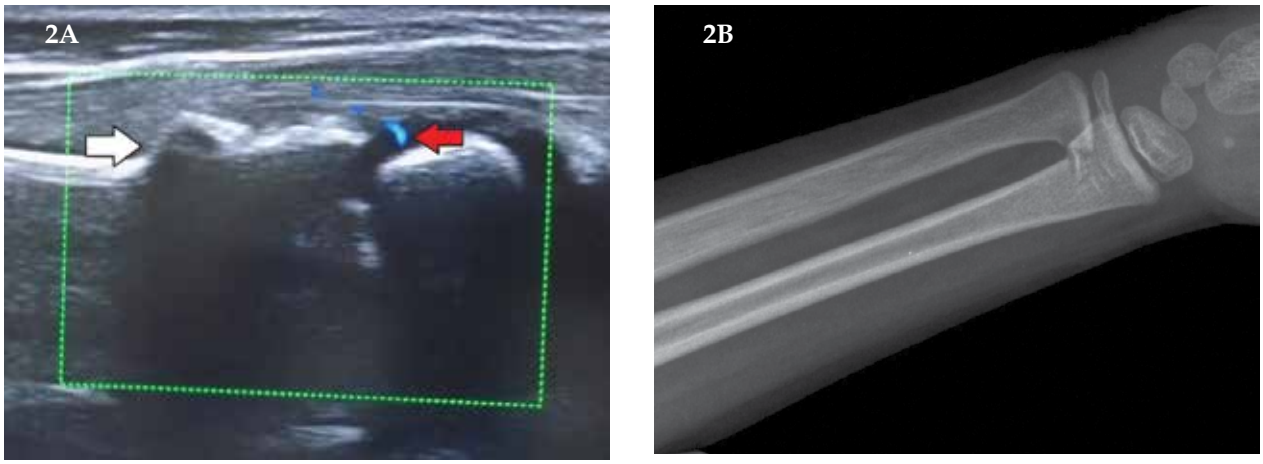

Fig. 2a. *Ultrasound picture of a buckle fracture (white arrow) and the nutrient vessel (red arrow).* ***2b****. X-Rays of the above mentioned fracture*

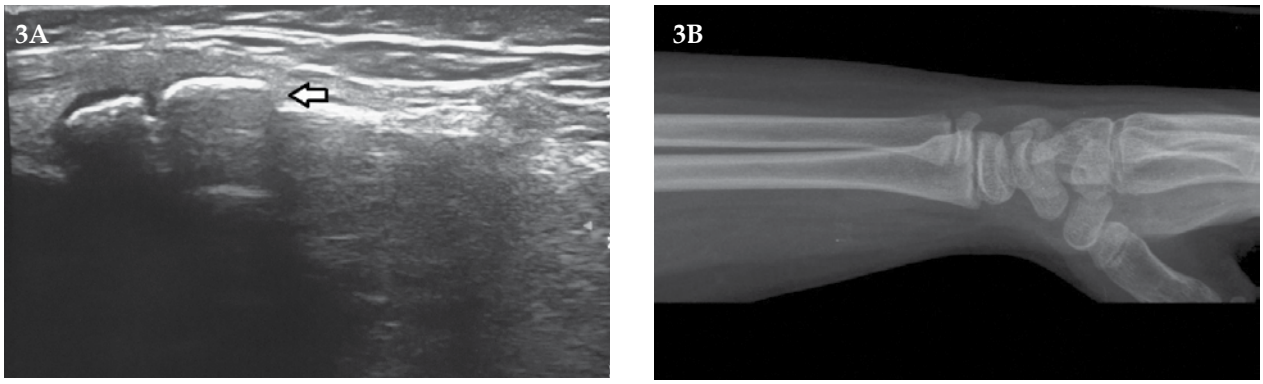

Fig. 3. *A buckle type fracture*

4. Discussion

Minimally displaced distal radius fractures in children counts for a significant portion of the distal forearm fractures [3].

To the author's knowledge the true annual incidence of such fractures is not known in Greece, however in the United Kingdom, 900,000 children attend A&E Department suffering of a distal forearm fracture [4, 5].

According the literature the sensitivity of ultrasound in detecting the minimally displaced distal forearm fractures is quite high [6] even higher than x-rays [5]. The results from the above study showed that ultrasound demonstrated the same sensitivity with X-rays. The ultrasonography can be also used to asses callus formation and fracture healing [5].

Ultrasonography is operator dependent, but detecting a fracture has a lower learning curve when compared to other more complicated soft tissue pathology. Additionally, the cost of an ultrasound is low and comparable to that of an X-ray. Finally, the use of ultrasound for the diagnosis of such a common but occult fracture saves children from radiation exposure.

The clinical implication of this study could be the use of a portable ultrasound device by the caring physician, in a primary care center could lead to a further cutdown of the direct cost of expensive X-rays equipment and staff. Furthermore, the indirect cost from waiting time at the A&E department, travelling from a primary care center to a referral hospital, lost working hours form accompanying persons, is also reduced. Conclusively, the application for ultrasonography could substantially contribute to the reduction of the direct and indirect health costs in fractures diagnosis.

5. Conclusions

The results showing that ultrasound can be safely

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used in diagnosis of minimally displaced distal radius fracture in children. It is a valuable bedside test, a low cost, safe and effective diagnostic tool and as sensitive as X-rays for the diagnosis of minimally displaced distal radius fractures in children.

Conflict of interest:
The authors declared no conflicts of interest.

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ΠΕΡΙΛΗΨΗ

Σκοπός: Ο σκοπός της παρούσας μελέτης, είναι η σύγκριση του ακτινολογικού με τον υπερηχογραφικό έλεγχο στη διάγνωση των ελάχιστα παρεκτοπισμένων καταγμάτων του κάτω πέρατος κερκίδας στα παιδιά. Παρουσιάζονται τα πρώιμα αποτελέσματα σε ένα επαρχιακό νοσοκομείο

Ασθενείς και Μέθοδος: Είκοσι τέσσερα παιδιά ηλικίας δύο έως δέκα τεσσάρων ετών, με κλινικά πιθανό κάταγμα κάτω πέρατος κερκίδος, υποβλήθηκαν σε υπερηχογραφικό έλεγχο. Ο ηχοβολέας τοποθετείτο αρχικά στην περιοχή της μέγιστης ευαισθησίας στην ψηλάφηση και ακολούθως η εξέταση επεκτείνετο κυκλοτερώς του οστού.

Αποτελέσματα: Σε όλες τις περιπτώσεις καταγμάτων υπήρχε ευαισθησία στην ψηλάφηση, ενώ όπου ετίθετο διάγνωση κατάγματος από τον ακτινολογικό έλεγχο, συμφωνούσε και η υπερηχογραφική διάγνωση.

Συμπεράσματα: Από την παραπάνω μελέτη, φαίνεται ότι ο υπέρηχος έχει τουλάχιστον την ίδια διαγνωστική αξία με τον ακτινολογικό έλεγχο, στα ελάχιστα παρεκτοπισμένα κατάγματα του κάτω πέρατος της κερκίδας στα παιδιά. Με την απλή και οικονομική αυτή απεικονιστική μέθοδο αποφεύγεται η έκθεση σε ακτινοβολία των νεαρών ασθενών.

ΛΕΞΕΙΣ ΚΛΕΙΔΙΑ: κατάγματα περιφερικής κερκίδας, παιδιά, υπέρηχος

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Management of children with congenital hypoplasia - defficient femur

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ABSTRACT

Congenital femoral hypoplasia is an extremely rare and severe dysplasia. The main characteristic is the severe leg length discrepancy. It covers a wide spectrum of femoral deformities ranging from hypoplasia to severe proximal deficiency. The aim of our study is to describe the management of the femoral deficiency. We used distraction histogenesis, with a circular Ilizarov frame, for the children that have prospective to equalize the discrepancy. A series of 6 children (5 girls and 1 boy) were treated over a period of 10 years. Patients were classified with the Pappas method. During the first 5-6 years of age, using appropriate orthotic devices, all of them had achieved a reasonable independence for their daily and school activities. In 3 of our patients treatment provided was distraction histogenesis using Ilizarov frame. For 1 patient, with untreatable discrepancy, we continue with special orthotic device. The increase in length was 5 cm to 6 cm, with an increase of 32% of the initial length (ranging from 33% up to 42%). The mean healing index was 17 days/cm. Severe complications presented during the procedures. Twice a child had fractures of the regenerated bone, soon after the removal of the device. They were treated with reapplication of the frame until union of the fractured bone. One child had knee subluxation that is partly corrected with realignment of the device at the knee joint. Treating children with deficient femur is a great challenge for the surgeon. It is almost impossible to correct in one procedure the whole discrepancy. The active life of the surgeon is not sufficient to correct all the deformities of the growing child.

KEY WORDS: congenital short femur; femoral deficiency in children; leg lengthening with Ilizarov

1. Introduction

Congenital hypoplasia of the femur is an extremely rare congenital disorder, with a reported incidence 1 per 100,000 newborns. The term femoral deficient child, similar with congenital hypoplasia, covers the spectrum of dysplastic femur, from simple hypoplasia to proximal femoral focal deficiency. In femoral hypoplasia, we refer in severe shortening of the femur up to

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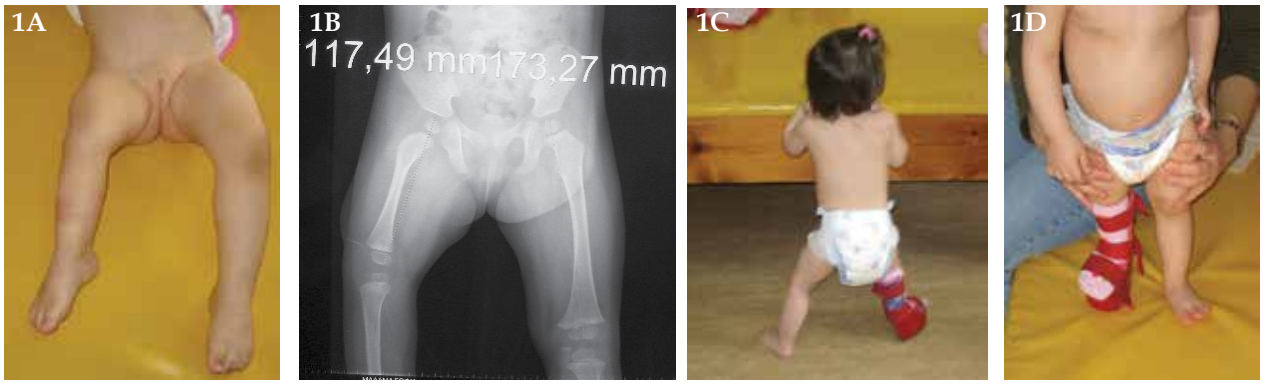

Fig. 1 a, b, c, d. *This girl, using appropriate orthotics and physiotherapy, achieved standing and walking without delay in her motor development*

10-40% of the length of the normal femur. Initial LLD is greater than 4 cm. All the femoral elements are present, but the femur is shorter and thinner than the normal side. The radiological examination reveals the hypoplastic femur, with the presence of the proximal and distal part of the femur [1, 2, 3].

In proximal femoral focal deficiency (PFFD), there is a spectrum of pathology of the proximal part of the femur, from partial shortening of the subtrochanteric area, to complete absence of the upper part, with severe shortening of the femur.

The lesion can be unilateral or bilateral [4]. There is a characteristic clinical picture with the affected leg in flexion, abduction, and external rotation. There is a variety of other lesions of the affected leg, as valgus knee, knee instability because of the absence of cruciate ligaments, fibular hemimelia, foot dysplasia like club foot or ball joint of the ankle.

The main feature of these children is the severe leg length discrepancy (LLD).

The use of distraction osteogenesis (Ilizarov) has completely changed the natural history of children with congenital femoral hypoplasia. It requires series of lengthening procedures, as it is difficult to equalize severe LLD in one procedure. Lengthening for more than 20% of the initial length has a lot of severe complications, but the final result is very encouraging [5, 6]. When the initial LLD is over the 50% of the length of the non affected femur, lengthening is difficult to achieve the purpose of equalization. The Van Nes procedure, using the ankle joint as a knee joint and fitting an appropriate orthotic, is the most suitable solution.

The purpose of our paper is firstly to present a series of 6 children with congenital femoral hypoplasia and secondly describe their management. The use of distraction osteogenesis is a challenge for the pediatric orthopaedic surgeon.

2. Patients and method

This is a series of children with femoral deficiency that are under our supervision since they are born. They are classified according to Pappas criteria in type 9 two girls, type 7 two girls, type 3 one boy and one girl with bilateral lesion.

Early in life, using appropriate orthotics and physiotherapy, all our children achieved standing and walking, without delay in the motor development. (**Fig. 1a, b, c, and d)**

From the age of 5 yrs, we started treatment of severe shortening of the leg. In order to start the plan of lengthening, there must be the option to equalize the leg. Using the multiplier charts of Paley [7], we estimated the final LLD at 12, 15, 16 , 22 cm and at the child with Pappas 3 more than 30 cm.

We could not estimate the final difference in the child with the bilateral affected femurs.

In 3 girls (type 9, and 2 of those with 7 Pappas classification) we used the leg lengthening procedure with the Ilizarov method, while for the child with the type 3 Pappas, we decided to proceed with orthotics, since equalization was not possible. In that child the initial position of the foot was at the level of the knee of the non affected limb.

One child is scheduled for lengthening during the

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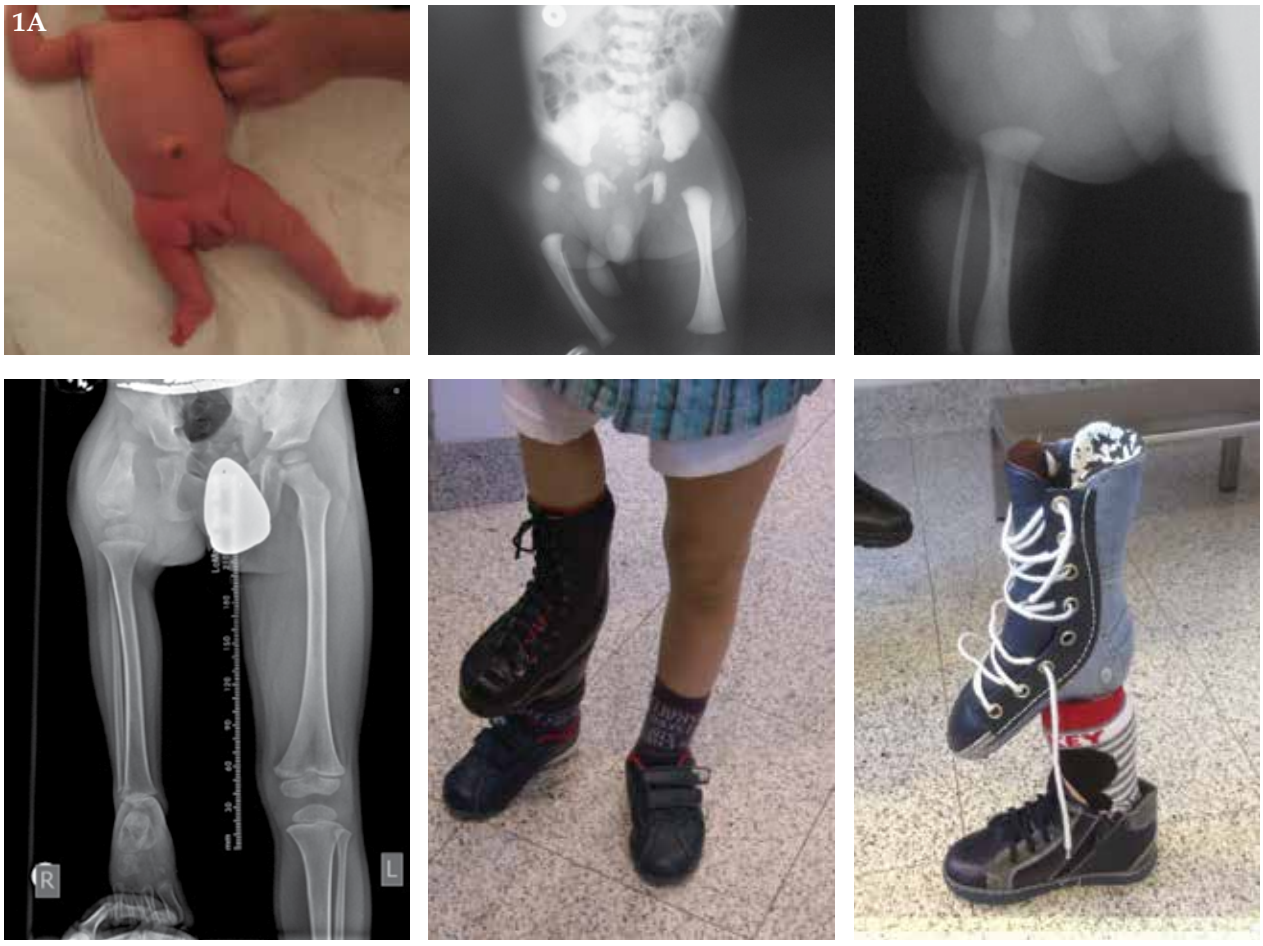

Fig. 2 a, b, c, d, e, f. *Clinical and radiographic features of a boy with untreatable discrepancy. He is planned to accept a Van Nes procedure in the future*

next year. The girl with bilateral affected femur, after the initial treatment and mobilization, had chosen to receive medical management elsewhere.

The boy with untreatable discrepancy had a special orthotic shoe, with double base, so he can walk independently. Further plan is to convert the ankle joint in knee joint (Van Nes procedure), once it will be required. (**Fig. 2a, b, c, Fig. 2d, e, and f)**

Three patients were treated with distraction histogenesis for the management of leg length discrepancy. One had initially treatment of the dislocated hip with closed management. We used a preassembly device, with circular frames that was constructed the previous days of the surgery. The system consisted from 3 rings. Stabilization of the knee was done during the progress of the lengthening. We did not use rings for the pelvis.

3. Results

One girl had a complete equalization of the limbs with an increase of the length of the femur for 5 cm, a 33% of the initial length of the affected femur, with a healing index of 17 days/cm. During the first days after the removal of the device, we noticed a small plastic deformation of the regenerate that was treated conservatively with non weight bearing and a functional brace of the femur. With 2 years follow up, the LLD is today 2cm, with a valgus knee (it was part of the initial dysplasia). Our plan is to perform a new procedure for leg lengthening at the age of 12, with epiphysiodesis with 8 plates for the valgus knee.

The 2 girls with initial LLD of 8 cm of the femur and 2 and 3 cm consecutively of the tibia, were treated with distraction histogenesis of the femur, gaining a lengthening of 35% and 42% of the length of the af-

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fected femur. They had a 6 cm increase of the femur.

All 3 girls had a nice consolidation of the regenerated femur. Our rate was increase of 0,1 cm per day in 4 daily intervals.

When we removed the device in the second girl, in 3 days time she had fracture of the regenerate. We reapplied the Ilizarov device to stabilize the fracture. The fracture healed but that was hard for the emotions of the girl. We achieved to reduce the LLD at 2 cm (there was also another 2 cm difference in the tibia). The same girl had a new lengthening procedure 2 years later, with an increase of 4 cm, but again at the removal of the device she had a fracture of the regenerate. We reapplied the Ilizarov device and achieved the healing of the fracture. Today, 1 year after her 2nd elongation, she is scheduled for a new procedure, since there is a LLD of 5 cm in the femur (there is a continuous increase in the LLD).

The last girl with an enormous increase of the length of the affected femur (43% of the initial length), was complicated from knee subluxation, that was partially treated with changes in the Ilizarov device, with hinges acting in the knee joint. Our patient, 2 years after the initial procedure of the lengthening of the femur and lengthening of the tibia, had a LLD of 5 cm, with varus deformation of the hip joint. Our plan is to correct the hip, perform a new lengthening of the femur and treat her equinus of the ankle that partially covers today the severe LLD.

4. Discussion

There are few series in the literature for the management of an extremely rare condition, like the femoral deficient child. Various types of the hypoplasia have been described.

Pappas classification has 9 types, with type 1 the most severe with complete absence of the proximal part and type 9 the less severe, where there is LLD and mild deformity in the subtrochanteric area [8].

Classification of Lloyd Roberts and Fixsen in 3 types was based on the hip stability, as the result of the absent part of the proximal femur. Aitken classification has 4 types, depending on the presence of the anlage of the femoral head. It is a guide for the type of surgical management of the problem. Paley has proposed a classification based on the stability of the hip and knee joint, focusing to elements that influence the results of leg lengthening procedures [9, 10, 11, 12].

We have chosen the Pappas classification as it has a more detailed description of the elements of the hypoplasia, both for the upper part as well for the whole femur.

Management of these children starts from the period they are babies, for the proper motor development. We must offer early physio assistance and appropriate orthotics for the standing and walking progress of the babies.

Appropriate orthotics that equalize the length discrepancy, permit children to function well for their motor development.

It is not possible to achieve equalization in all cases of femoral deficiency. If the initial discrepancy is more than 50% of the length of the normal leg or if the foot of the affected limb is at the level of the knee joint of the other side, then the continuous developing discrepancy cannot be covered with continuous limb lengthening procedures. The need for continuous severe procedures until the adult life, makes the choice of using the ankle joint as a knee joint (Van Nes procedure) the most appropriate one [13,14].

Lengthening a hypoplastic femur is an extremely difficult procedure, since we treat both the hypoplastic bone and the stiff shortened muscles. It requires continuous monitoring during the lengthening [5, 6, 15].

In our series the regenerate had a good radiological consolidation and we had not cases to change the rate of lengthening. It is possible, when we lengthen already lengthened femurs, to have weak regenerate. Then we may delay the rate of lengthening.

The rate of complications when lengthening hypoplastic femur is extremely high. In the literature it is been referred that there is almost 100% fracture rate of the newly formed bone, after removal of the external device. Using an intramedullary pin at the beginning of the procedure is essential for the protection of the new bone. Using today intramedullary lengthening devices or submuscular plates, the incidence of fractures has been severely reduced. For our cases, with fractures of the new bone, we reapplied the Ilizarov device. In our next

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case, we plan to use, from the beginning, an intramedullary pin [16 -24].

A common complication when we lengthen the femur is the dislocation or subluxation of the knee joint. It is the result of the shortened posterior thigh muscles in combination with the absence of cruciate ligaments and hypoplasia of the femoral condyles. This complication is common when the lengthening is more than 20-25% of the initial length. In our case, this happened because we overestimated our abilities, performing lengthening of 42% of the initial length. Using a ring to stabilize the knee in extension or special joints for controlled knee motion, can protect the knee from subluxation. The same complication exists for the hip joint, a common complication also. It requires regular radiological evaluation, mainly when the lengthening is exceeding 5-6 cm [5, 6, 25, 26].

In conclusion, management of children with deficient femur is a great challenge for the orthopaedic surgeon. Trying to restore as much as possible, severe leg length discrepancies, we overestimate our abilities for lengthening of a small hypoplastic limb. These procedures start from a baby and end up for the patient as an adult. There is a very strong emotional relation between the surgeon and the child and his family. We must plan with an accurate time table the whole management, including our procedures in the school activities of these children.

The active life of an orthopaedic surgeon is not sufficient enough to deal all these problems of the growing child, as underlined always G Pistevos, the pioneer of lengthening procedures in children in Greece.

Conflict of interest:

The authors declared no conflicts of interest.

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ΠΕΡΙΛΗΨΗ

Η συγγενής υποπλασία του μηριαίου αποτελεί μια βαρύτατη και ιδιαίτερα σπάνια διαμαρτία, με κύριο χαρακτηριστικό την μεγάλη ανισοσκελία. Καλύπτει μεγάλο εύρος διαμαρτιών του μηριαίου, από υποπλασία έως απουσία του εγγύς τμήματος. Σκοπός της εργασίας μας είναι να παρουσιάσουμε την αντιμετώπιση παιδιών με υπολειμματικό μηριαίο. Χρησιμοποιήσαμε την διατατική ιστογένεση, με την συσκευή Ilizarov, στα παιδιά που είχαν προοπτική να ισοσκελισθούν.

Παρουσιάζουμε μια σειρά από έξι 6 παιδιά που αντιμετωπίζουμε από την γέννησή τους. Πρόκειται για 5 κορίτσια και 1 αγόρι. Η κατάταξή τους έγινε με τα κριτήρια Pappas.

Με την χρήση ειδικής κατασκευής ορθωτικών για την κάλυψη της ανισοσκελίας, επιτεύχθηκε η αυτόνομη βάδιση, χωρίς να υπάρχει σημαντική κινητική υστέρηση. Από την ηλικία των 5 ετών, σε 3 παιδιά, έγινε η διατατική ιστογένεση με την μέθοδο Ilizarov, ενώ σε 1 παιδί κρίθηκε μη εφικτή και αντιμετωπίζεται συντηρητικά. Μια ασθενής θα αντιμετωπισθεί στο μέλλον ενώ μία ασθενής αντιμετωπίζεται αλλαχού.

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Η επιμήκυνση ήταν από 5-6 εκ, με επίτευξη αύξησης του μήκους από 33 έως 42% του αρχικού μήκους. Ο μέσος όρος του δείκτη ωρίμανσης ήταν 17 ημέρες ανά εκατοστό.

Εμφανίσθηκαν πολλαπλές επιπλοκές στην πορεία της επιμήκυνσης. Αντιμετωπίσαμε κάταγμα του νεοσχηματισθέντος οστού, αμέσως μετά την αφαίρεση της συσκευής. Εφαρμόσθηκε εκ νέου σύστημα Ilizarov για την πώρωση του κατάγματος. Μία ασθενής με υπεξάρθρημα του γόνατος αντιμετωπίσθηκε με ειδική αρθρούμενη εφαρμογή του συστήματος στο γόνατο, με μερική διόρθωση της βλάβης.

Παρουσιάζουμε τις ιδιαίτερες δυσκολίες στην εφαρμογή της διατατικής ιστογένεσης λόγω του μικρού αρχικού μήκους του μηρού και δράσης των ρικνών μυών.

Η αντιμετώπιση της υποπλασίας του μηρού αποτελεί μια πρόκληση για τον ορθοπαιδικό. Πρόκειται για ιδιαίτερα δύσκολο πρόβλημα και με πολλαπλές επιπλοκές στην πορεία αλλά με εντυπωσιακά αποτελέσματα στο τέλος της θεραπείας. Η ενεργός ζωή του Ορθοπαιδικού δεν είναι επαρκής για την αντιμετώπιση όλων των προβλημάτων του αναπτυσσόμενου παιδιού.

ΛΕΞΕΙΣ ΚΛΕΙΔΙΑ: συγγενής υποπλασία μηριαίου, ανεπάρκεια μηριαίου παιδιών, επιμήκυνση μηριαίου Ilizarov