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1- Persistent torticollis, facial asymmetry, grooved tongue, and dolicho-odontoid process in connection with atlas malformation complex in three family subjects

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Eur Spine J 2007 Dec;16 Suppl 3:265-70.
[Case report - Article in English]

Congenital clefts and other malformations of the atlas are incidental findings identified while investigating the cervical spine following trauma. A persistent bifid anterior and posterior arch of the atlas beyond the age of 3-4 years is observed in skeletal dysplasias, Goldenhar syndrome, Conradi syndrome, and Down's syndrome. There is a high incidence of both anterior and posterior spina bifida of the atlas in patients with metabolic disorders, such as Morquio's syndrome [Baraitser and Winter in London dysmorphology database, Oxford University Press, 2005; Torriani, Lourenco in Rev Hosp Clin Fac Med Sao Paulo 53: 73-76, 2002]. We report two siblings and their mother, with congenital, persistent torticollis, plagiocephaly, facial asymmetry, grooved tongues, and asymptomatic «dolicho-odontoid process». All are of normal intelligence. No associated Neurological dysfunction, paresis, apnoea, or failures to thrive were encountered. Radiographs of the cervical spine were non-contributory, but 3D CT scanning of this area allowed further visualisation of the cervico-cranial malformation complex in this family and might possibly explain the sudden early juvenile mortality. Agenesis of the posterior arch of the atlas and bifidity/clefting of anterior arch of the atlas associated with asymptomatic «dolicho-odontoid process» were the hallmark in the proband and his female sibling. Some of the features were present in the mother. All the family subjects were investigated. To the best of our knowledge the constellation of malformation complex in this family has not been previously reported.

2- Traumatic hip dislocations in children

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Rev Chir Orthop 2008 Feb;94(1):19-25.
[Original Article - Article in French]

PURPOSE OF THE STUDY

Traumatic hip dislocation is a rare event in children. Appropriate management remains a subject of debate. The purpose of this study was to investigate the epidemiological, therapeutic features of this situation and the long-term outcome after treatment.

MATERIAL AND METHODS

This was a retrospective analysis of 15 traumatic hip dislocations collected over a period of 20 years in pediatric patients with at least two years follow-up. We searched
for predisposing factors and factors affecting prognosis.

RESULTS

The series included 11 boys and three girls, mean age eight years. Dislocation was posterior in 13 hips and anterior in two. Time to reduction was less than 3h in eight cases, 3-6h in five and greater than 6h in two. After reduction, traction was performed in nine children, for 20 days on average, followed in five cases by immobilization for 40 days on average. Five hips were immobilized directly after reduction. We identified two groups by age: group 1 with dislocations in children aged less than six years (seven children) were characterized by low-energy trauma. Dislocation was not associated with other lesions. Predisposing factors (overt ligament hyperlaxity, insufficient superolateral head cover, coxa valga) were noted in six children. Reduction was simple. Later treatment consisted in immobilization with a pelvispedious cast for 30-45 days. Group 2 were dislocations in children aged over six years (seven children) victims of high-energy trauma. Associated injuries were frequent. Predisposing factors were not present. At mean 11 years follow-up, all hips are considered normal clinically. The radiograph was normal for 14 hips. In one case, there was a slight coax magna. In three patients, defective femoral head cover persisted. Coxa valga persisted in two patients.

DISCUSSION

Traumatic dislocation of the hip joint is rare in very young children, but results from a minimally traumatic event. This suggests the presence of predisposing factors in this category of patients, particularly capsulo-ligamentary hyperlaxity. After reduction, immobilization can be recommended. Traumatic hip dislocations in children are different from the adult variety due to their rarity, the general absence of associated fractures, easy reduction and better prognosis. The epidemiological and therapeutic features in children older than six years are however similar to those in adults.

3- Acute hematogenous osteomyelitis of the neck of the femur in children: 28 cases

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Rev Chir Orthop 2008 Feb;94(1):49-57.
[Original Article - Article in French]

PURPOSE OF THE STUDY

Osteomyelitis of the neck of the femur is uncommon, often with a misleading clinical presentation. We report a series of 28 cases of osteomyelitis of the femoral neck to illustrate the particular clinical and imaging findings related to this localization.

MATERIAL AND METHODS

This was a series of 28 children treated in our unit from 1990 through 2004: 17 boys and 11 girls, mean age eight years (range one month to 14 years). We analyzed the data in this series using a standard checklist, which noted the diagnostic and therapeutic measures. Results were analyzed by studying the complications, anatomic and functional outcome at mean follow-up of 3.5 years.

RESULTS

Time from symptom onset to consultation was five days on average with a delay of 4.5 days from consultation to hospitalization. Intense pain was noted for eight patients (28%) and total functional incapacity of the limb was noted for 15 (53%). Hip stiffness was observed in 11 patients (39%). The diagnosis of osteomyelitis of the femoral neck was established on the basis of imaging (MRI or scintigraphy) in three patients with bacteriological proof in two, of operative findings which confirmed the neck localization in 19, and on changes in the X-ray image of the neck in six. A positive bacteriology was noted in 71%. The germ was isolated from blood cultures and local samples obtained by arthrotomy in five patients (same germ), only in blood cultures for four, and only in local samples in 11. Met-S Staphylococcus aureus was isolated in 18 patients, Met-R S. aureus in one and a Streptococcus in one. All patients were given medical treatment and 25 underwent surgery. There were five thromboembolic complications and five patients who developed femoral pandiaphysitis. Results were analyzed at mean 3.5 years (range four months to 14 years). Complete hip motion was recovered in 78%. There were four cases of hip stiffness and two cases of ankylosis. Partial cephalic necrosis was noted in two hips and total necrosis of the head and neck in two.

DISCUSSION

Little data is available in the literature on isolated osteomyelitis of the femoral neck. Based on the pathogenic mechanisms known for osteomyelitis, an isolated localization in the neck of the femur, with no other site in the hip joint, is quite possible in an early stage of infection. We discuss the specific clinical and imaging features of this localization. Analysis of our findings show that the prognosis of femoral neck osteomyelitis is directly related to time to management. Outcome is poorer when treatment is started late. Prognosis is poor if pandiaphysitis develops.

4- Sclerosing epithelioid fibrosarcoma. A case report

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Rev Chir Orthop 2008 Feb;94(1):92-5.
[Case Report - Article in French]

Sclerosing epithelioid fibrosarcoma is a rare variant of fibrosarcomas, which was recently identified as a separate entity due to specific histologic and immunohistochemistry features and its poor prognosis. We report a case of sclerosing epithelioid fibrosarcoma of soft tissues, which developed in a 37-year-old woman who presented a tumor involving the posteromedial aspect of the left knee and which progressed in size for one year. Imaging revealed a well-delimited tumor process measuring 8 cm in its largest diameter and situated in the medial compartment of the left knee. Histology of the tumorectomy specimen and the immunohistochemistry study led to the diagnosis of sclerosing epithelioid fibrosarcoma of soft tissues. This new case illustrates the characteristic features of this tumor and recalls the difficult pathological diagnosis.

5- Osteoid osteoma of the talus and simple bone cyst of the homolateral calcaneus. A rare association

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Rev Chir Orthop 2008 Feb;94(1):96-9.
[Case Report - Article in French]

Osteoid osteoma and simple bone cyst are readily observed in long bones, but are much less common in the short bones of the foot. The association of these two tumors in the same foot is exceptional. We report the case

of a 15-year-old girl who presented an osteoid osteoma of the talus and a simple bone cyst of the calcaneus of the left foot. The patient complained of pain in the rear foot for four months which worsened at night and was of the inflammatory type. The physical examination was normal. Standard x-rays revealed a simple bone cyst in the calcaneus which could not explain the nighttime inflammatory pain. Scintigraphy and computed tomography of the left foot revealed an osteoid oseoma of the talus. Biopsy total resection of the osteoid osteoma with curettage and filling with cancellous bone of the simple bone cyst were performed. The patient has been free of recurrence at three years follow-up.

6- Results of surgical treatment of congenital convex pes valgus (10 non-idiopathic feet)

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Rev Chir Orthop 2008 Apr;94(2):128-34.
[Original Article - Article in French]

PURPOSE OF THE STUDY

Congenital pes valgus is a rare and complex deformity of the foot raising serious diagnostic and therapeutic challenges. The purpose of our work was to present the surgical procedures used in our series and to analyse outcome.

MATERIAL AND METHODS

Ten feet presenting congenital convex valgus treated surgically over a six-year period using the same operative technique were reviewed at minimum five years follow-up. Idiopathic deformities were excluded from this series. Deformities were secondary to arthrogryposis in five feet, a multiple malformative syndrome in four and diastematomyelia in one. The surgical technique used two approaches: a posteromedial incision to release the dorsal flexors, disinsert the tibialis posterior, open the talonavicular joint, release the Achilles tendon and release the posterior tibiotalar capsule; a lateral incision to lengthen the fibular tendons and perform an osteotomy of the anterior process of the calcaneum. A talonavicular pin and a calcaneocuboid pin maintained the correction. The tibialis posterior tendon was reinserted on the anterior aspect of the talonavicular capsule after incision of the dislocation chamber.

RESULTS

Outcome was considered good in five cases and fair in five. Outcome was fair in the arthrogyrposis feet. Undercorrection was observed in two feet and valgus flatfoot in three. Talar necrosis occurred in one foot and navicular necrosis in two.

DISCUSSION

Simultaneous correction of the different anomalies observed in the congenital convex foot was achieved in this series. The anatomic and functional results were satisfactory. We recommend avoiding overly extensive release in order to decrease the risk of talar and navicular necrosis. It is also important to check the reduction radiographically during the operation. Patients should use an orthesis several months postoperatively to avoid recurrence.

7- Acute hematogenous osteomyelitis of the obturator rim in seven children

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Rev Chir Orthop 2008 Apr;94(2):168-173.
[Original Article - Article in French]

PURPOSE OF THE STUDY

Acute osteomyelitis of the rim of the obturator foramen is rarely reported. The clinical presentation is atypical, making diagnosis a difficult task. We report a series of seven cases of osteomyelitis of the obturator rim which illustrate the different features of this localization.

MATERIAL AND METHODS

The series included seven boys with osteomyelitis involving the obturator foramen. We reviewed the clinical history, the diagnostic approach and management. Complications and anatomic outcome were noted at mean three years follow-up (range one to 10 years).

RESULTS

Mean age was 9.5 years (five to 12 years). Bilateral involvement was noted in one boy. Mean time from symptom onset to consultation was five days and mean time from consultation to hospitalization was eight days. No specific clinical presentation could be identified. Fever was not a constant feature. Mean body temperature was 38.6 degrees C and was not greater than 38.5 degrees C in four children. Symptoms were limited to hip pain in five cases and abdominopelvic pain was noted in two. Physical examination failed to trigger exquisite ischial or pubic pain in two patients. The osteomyelitis involved the ischiopubic ramus in four cases, the ischium in two with one bilateral case, and the pubis in one. Certain diagnosis was established as follows: MRI findings plus isolation of the pathogenic agent (n=4) ; the plain x-ray showed a defect in the ischiopubic ramus, MRI showed signs favoring osteomyelitis of the ischiopubic ramus and surgery evacuated a purulent collection (n=1) ; strong uptake of the obturator rim on scintigraphy (n=2). Medical treatment was given in all cases and surgery was performed in six patients. Outcome was noted at three years follow-up, range one to 10 years. All patients recovered normal function. There were no complications.

DISCUSSION

Hematogenous osteomyelitis of the pelvis is exceptional (2-11% of cases of osteomyelitis). Localization in the obturator rim is rare. We discuss the specific diagnostic

and therapeutic features of this localization. Despite the controversy concerning the treatment of pelvic osteomyelitis and in particular cases involving the obturator rim, prognosis is generally good. No complications have been described in the literature specifically associated with osteomyelitis of the obturator rim.

8- Desmoplastic fibroblastoma of the foot

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Rev Chir Orthop 2008 Apr;94(2):188-192.
[Case Report - Article in French]

Desmoplastic fibroblastoma is a benign, rare, slow-growing soft tissue tumor which is found in a wide anatomic distribution, predominantly in adult males. The characteristic gross aspect is that of a typical cartilage-like tumor which histologically presents regular fibroblastic proliferation, often in a stellar shape within a dense collagen or myxo-collagen stroma. We report a new case observed in the foot and study the anatomic and clinical aspects of this rare entity.

9- Fibrous dysplasia of the rib. Ten case reports

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Rev Chir Orthop 2008 May;94(3):301-307.
[Case Report - Article in French]

PURPOSE OF THE STUDY

Fibrous dyplasia is a rare sporadic disease accounting for 0.8% of primary bone tumors. This benign pseudotumor results from proliferation of fibrous tissue in bone and the production of immature bone tissue without an osteoblastic crown. The disease can involve one or more bones, ribs are rarely involved. We report a series of 10 cases of costal fibrous dysplasia.

CASE REPORTS

This series of 10 cases was collected over a period of 10 years (1996-2005). There were five men and five women, mean age 38.4 years, range 27-52 years. One rib was involved in eight patients, two ribs in two. Pain was the most frequent symptom. Plain X-rays showed signs suggestive of fibrous dysplasia. To confirm the diagnosis, rib resection was performed in all ten patients. Pathology examination provided the diagnosis. The postoperative period was uneventful in all patients and all are recurrence free at mean 50 months follow-up.

DISCUSSION

Management of fibrous dysplasia can be simple surveillance in the majority of patients. Nevertheless, in the event of a single focus, particularly in a rib, fibrous dysplasia can raise a difficult problem of differential diagnosis with malignant tumors. Surgical resection is therefore required in selected cases.

10- Early results of the Ponseti method using the Steenbek foot abduction brace: a prospective study of 95 feet

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J Pediatr Orthop B. 2008 May;17(3):134-8.
[Original Article - Article in English]

The purpose of this study is to evaluate the early results of the Ponseti method and the effectiveness of the Steenbek foot abduction brace. A total of 74 patients with 110 idiopathic clubfeet were included in this prospective study. The feet were evaluated according to the Dimeglio-Bensahel classification, the Catteral-Pirani classification and the functional classification of the Hospital for Joint Diseases. Ninety-eight feet (89%) had a good result after the casting period. All the feet evaluated after the period of full-time bracing and during the period of part-time bracing showed a good correction. The Ponseti method using the Steenbek foot abduction brace is effective in correcting idiopathic clubfeet.

11- A hypoplastic atlas and long odontoid process in a girl manifesting phenotypic features resembling spondyloepimetaphyseal dysplasia joint laxity syndrome

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Skeletal Radiol. 2008 May;37(5):469-73.
[Case Report - Article in English]

Phenotypic features consistent but not completely diagnostic for spondyloepimetaphyseal dysplasia joint laxity (SEMDJL) were encountered in a 7-year-old-girl. Additional tomographic features of a hypoplastic atlas (assimilation of the posterior arch of the atlas) and unduly long odontoid process were seen. We report what might be a novel type of SEMDJL.