POSITIVE

Official newsletter of the Paediatric Orthopaedic Society of India

Editorial

India has the largest child population in the world. We as paediatric orthopaedic surgeons come across a wide variety of disorders. The management of these children generates a considerable amount of data which is not being disseminated. In the hour of need many a surgeon, in a non-institutional set up have difficulty accessing the full-text of relevant articles from specialist paediatric journals. All that is required is an Indian journal dedicated to Paediatric Orthopaedics through which the work done in our country and relevant to our practice can be published and is freely accessible. The POSITIVE in future is aimed to be an indexed journal. The scope of this journal would be original research articles, original clinical observations accompanied by analysis and discussion, analysis of philosophical, ethical, or social aspects, review articles, statistical compilations, technical tips, description of evaluation of methods or procedures, case reports with discussions and letters to the editors, all pertaining to paediatric orthopaedics. The journal is proposed to be published biannually on a regular basis. This requires participation and contribution from the Indian orthopaedic community, especially the members of Paediatric Orthopaedic society.

Dr William G Mackenzie, Dr Elizabeth A Szalay, Dr Dhiren Ganjwala and Dr Abhay Gahukamble have made valuable contributions to this issue. Dr Harish P, fellow in Paediatric Orthopaedics at CMC Vellore, has functioned as the assistant editor for this issue and his help is gratefully acknowledged.

Vrisha Madhuri.

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September 2009

POSI
***T*RANSFERS**
***I*NFORMATION**
***V*ERY**
***E*FFECTIVELY**

SHORT STATURE IN SKELETAL DYSPLASIA

William G Mackenzie

Short stature is a common presentation in paediatric orthopaedic practice. A diagnosis needs to be established for appropriate management.

INTRODUCTION

Short stature is defined as height less than two standard deviations from the mean for that population at a given age or height that is less than the third percentile for the chronological age of the child.

The term dwarf refers to disproportionate short stature while the term midget refers to proportionate short stature. The disproportion could be either due to short limbs or short trunk. The short-limb types are further categorised based on which segment of the limb is short. Rhizomelia refers to shortening of the root (proximal) portion of the limb, mesomelia to the middle segment, and acromelia to the distal segment. Some of these disorders are named after the morphology of the skeleton (diastrophic means to grow twisted, camptomelic means bent limbs and chondrodysplasia punctata refers to stippled cartilage). Eponyms are used to name other disorders (e.g., Kniest, Morquio, and McKusick).

Body proportion refers to the ratio of upper and lower segments. Upper segment extends from the top of the head to the pubic symphysis and lower segment extends from the symphysis pubis to the sole of the feet. The head and trunk grow faster than the extremities in early life; therefore the ratio which is 1.7 in infants reaches a value of 1.0 around 10 years of age.

ASSESSMENT

When parents present with a child who is shorter than other children of his age group or siblings, two important points to be considered for evaluation are to ascertain the age at onset of short stature and the presence of any similar family history. Occasionally the problem is detected prenatally on routine pre-natal ultrasound.

Other aspects of evaluation include skeletal features, short trunk/ short limb, extra skeletal features, radiographic features, laboratory tests and genetic analysis.

PRENATAL DIAGNOSIS

With the increasing use of routine prenatal screening, increasing number of patients with skeletal dysplasia are being diagnosed before birth. When ultrasound shows a foetus with shortening of the skeleton, femur length is the best biometric parameter for distinguishing among the five most common possible conditions.

- Foetuses with femur length less than 40% of the mean for gestational age have Achondrogenesis.
- Femur length between 40% and 60% imply Thanatophoric dysplasia or Osteogenesis imperfecta type II.
- Femur length over 80% implies Achondroplasia or Osteogenesis imperfecta type-III.

Further testing may be performed, if indicated, by chorionic villous sampling and mutation analysis. (Table 1)

AGE AT RECOGNITION

Skeletal Dysplasias can also be differentiated according to the age at which they are recognized. (Table 3)

William G Mackenzie, MD,
Chairman of Orthopaedics,
Alfred I. DuPont Hospital for Children,
Wilmington, Delaware, USA.

Table 1: Genetic Analysis

***FGFR3* group (local regulator of cartilage growth)**	Achondroplasia Hypochondroplasia Thanatophoric dysplasia
***COL1A* group (structural osseous protein)**	Osteogenesis imperfecta
***COL2A1* group (structural cartilage protein)**	SED Kniest dysplasia Stickler dysplasia Strudwick dysplasia SED tarda
Defective sulfate transport enzyme group	Diastrophic dysplasia Achondrogenesis
Collagen oligomeric matrix protein group (structural cartilage protein)	MED Pseudoachondroplasia
Storage disorders	Mucopolysaccharidoses Mucolipidoses

Table 2: Pattern of Inheritance

Age of Recognition	Dysplasia	
At Birth or in infancy	***Short Limb*** ▪ Achondroplasia ▪ Hypochondroplasia ▪ Diastrophic dysplasia ▪ Ellis van Creveld ▪ Metatropic dysplasia ▪ Chondrodysplasia punctata ▪ Achondrogenesis ▪ Thanatophoric dysplasia	***Short Trunk/limb*** ▪ Spondylo-epiphyseal dysplasia ▪ Kniest dysplasia
Those not recognizable at birth or in 1st year of life	▪ Storage Disorders -Morquio Syndrome ▪ Pseudoachondroplasia ▪ Multiple Epiphyseal dysplasia ▪ Metaphyseal dysplasia - McKusick, Schmidt	

Table 3: Age at Recognition

AUTOSOMAL DOMINANT	▪ Achondroplasia ▪ Hypochondroplasia ▪ Pseudoachondroplasia ▪ Multiple Epiphyseal dysplasia ▪ Spondyloepiphyseal dysplasia congenita ▪ Kniest dysplasia
AUTOSOMAL RECESSIVE	▪ Morquio's ▪ Ellis van Creveld ▪ Diastrophic dysplasia
X- LINKED	▪ SED tarda

SKELETAL FEATURES

Skull and face

- Skull and face are almost normal in Hypochondroplasia, Pseudoachondroplasia, Metaphyseal chondrodysplasia, and MED.
- Frontal bossing and midface hypoplasia is seen in Achondroplasia and Thanatophoric dysplasia.
- Abnormal teeth are present in Chondroectodermal dysplasia and Osteogenesis Imperfecta**.**
- Prominent cheeks, circumoral fullness (Cherub dwarf) and cauliflower ears are observed in Diastrophic dysplasia.
- Prominent eyes and forehead, depressed midface and cleft palate are seen in Kniest syndrome and SED.
- Hypertelorism, depressed nasal bridge and a bifid nasal tip are features of Chondrodysplasia punctata.

Short limbs

- Rhizomelia (Fig 1) is seen in Achondroplasia, Pseudoachondroplasia, Chondrodysplasia punctata and Diastrophic dysplasia; Mesomelia is seen in Leri-Weill dysplasia and Hypochondroplasia.

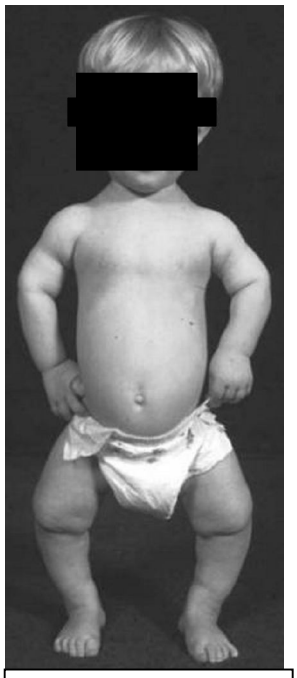


Fig 1: Rhizomelic dwarf

- Acromesomelia is seen in Chondroectodermal dysplasia.

Short trunk

Trunk is short in SED and Kniest dysplasia due to significant involvement of spine (Fig 2).

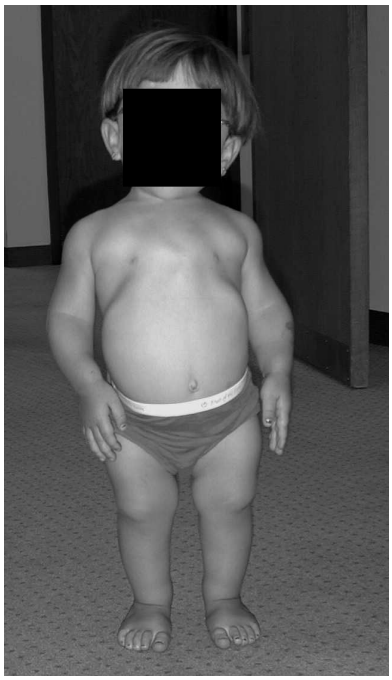


Fig 2: Short trunk and limb dwarf

Chest

Chest deformities are frequent in short stature. Pectus carinatum is seen in SED, Metatropic dwarfism and Morquio's Syndrome, whereas a depressed sternum is a feature of Kniest syndrome.

Spine

- A normal spine is present in Hypochondroplasia, MED and Chondroectodermal dysplasia.

- Thoracolumbar kyphosis, lumbar lordosis and scoliosis are seen in most of the skeletal dysplasias as is cervical instability which is present in most except in Achondroplasia. In Achondroplasia neural compression can still occur at the foramen magnum due to asynchronous development of neural elements and base of the skull.

- Multilevel spinal canal stenosis, common in the lumbar spine is present characteristically in Achondroplasia. Stenosis also occurs in Hypochondroplasia.

- Coccygeal tail (cartilaginous prolongation of the coccyx) is characteristic of Metatropic dysplasia.

Hands

- Shortening of middle finger and a gap between the third and fourth digit giving the appearance of a trident is seen in Achondroplasia.

- Short, broad, and ulnar deviated hands, Hitchhiker thumb and symphalangism are all features of Diastrophic dysplasia.

- Madelung deformity is a feature of Dyschondrosteosis (Leri–Weill).

- Polydactyly occurs in Chondroectodermal dysplasia.

Hips

- Coxa valga deformity is seen in Dyschondrosteosis (Leri–Weill) and Chondro-ectodermal dysplasia, whereas coxa vara is seen in Chondrodysplasia punctata, MED, SED and Kniest dysplasia.

- Coxa magna or flattening of femoral head is seen in SED and Diastrophic dysplasia.

Knees

Various knee deformities are seen. *(Fig 3)*

- Genu varum deformity occurs in Achondroplasia due to relatively long fibula. It is also observed in Hypochondroplasia, Metaphyseal dysplasia and Pseudoachondroplasia.

- Genu valgum is seen in SED, MED, Morquio's syndrome, Ellis van Creveld and Kniest dysplasia.

- In Pseudoachondroplasia both the deformities can occur because of associated ligamentous laxity. Characteristically, wind swept deformity of the knees is seen in these patients.

Most of the dysplasias have a propensity to develop early arthritic changes often in the large joints, i.e., the hips and the knees.

Foot and Ankle

- Varus deformity at the ankle is seen in Achondroplasia and Metaphyseal dysplasia.

- Ankle valgus is seen in MED and Dyschondrosteosis (Leri – Weill).

Club foot (predominantly equinovarus) occurs characteristically in Diastrophic dysplasia, Kniest and SED.

EXTRA SKELETAL FEATURES

Skull

Normotensive hydrocephalus, cervicomedullary compression causing developmental delay, hypotonia, sleep apnea, feeding difficulties, hemiplegia, quadriplegia and sudden death commonly occurs in Achondroplasia.

Eye

- Cataracts occur in storage disorders.
- Retinal detachment is observed in SED Congenita and Kniest dysplasia.

Ear

- Recurrent otitis media occurs in most of the skeletal dysplasia. It is common in Achondroplasia due to improper functioning of eustachian tubes. Hearing loss follows repeated infection. It may also be due to abnormal ossicles. Hearing loss is also a feature of Kniest dysplasia.

Cardiac system

- Congenital cardiac anomalies, commonly atrial septal defect or single atrium, with short stature and skeletal dysplasia are seen in Ellis van Creveld syndrome (Chondroectodermal dysplasia).

- Acquired cardiac disorders develop in storage disorders.

Respiratory system

- Laryngotracheomalacia occurs in Camptomelic dysplasia and Diastrophic dysplasia.

- Apnea, central or obstructive is seen in Achondroplasia.

- Restrictive lung disease (small, stiff thorax) develops in Metatropic, Thanatophoric & Ellis van Creveld dysplasias.

- Immunological diseases of lungs develop in McKusick metaphyseal chondrodysplasia.

RADIOGRAPHY

Radiographs which are done routinely are:

- Cervical spine- Flexion and extension lateral views to look for instability. An atlanto-dens index of more than 5 mm on flexion and extension lateral radiographs indicates instability.

- Standing AP and lateral radiographs of thoracolumbar spine to assess the spinal involvement. In Achondroplasia there is progressive narrowing of the interpedicular distance from proximal to distal especially at the lumbar level. Lateral view of the spine in Achondroplasia shows reduced distance between the vertebral body and lamina due to short pedicles.

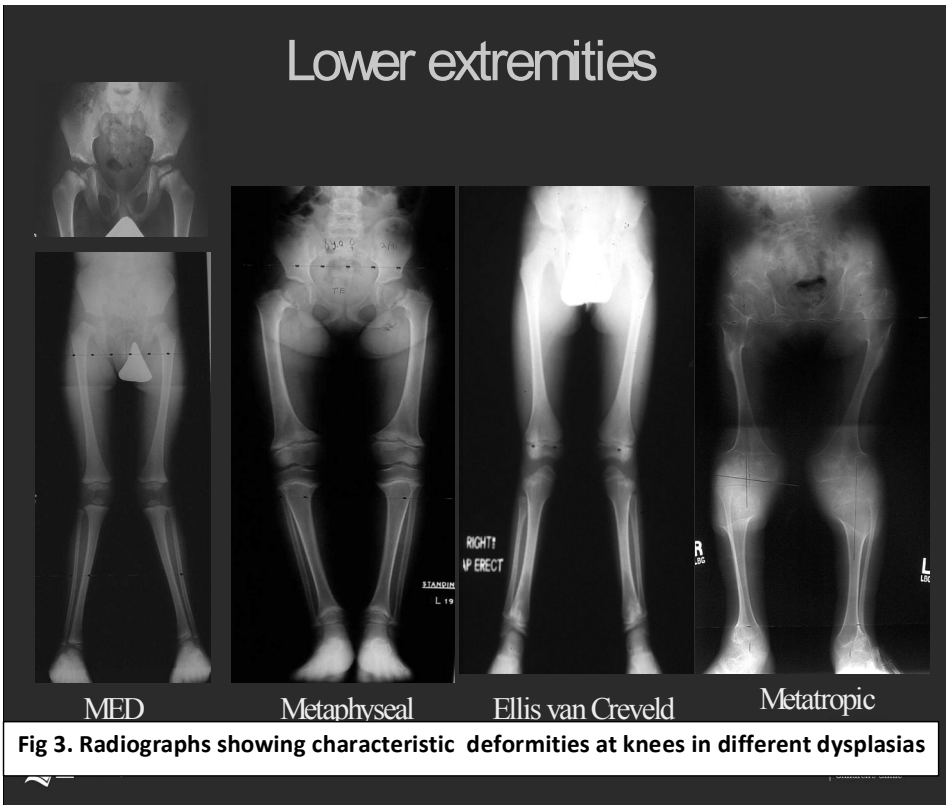


Fig 3. Radiographs showing characteristic deformities at knees in different dysplasias

- Standing AP and lateral views of both lower limbs from hip to ankle is required to assess alignment when deformity correction is planned.

- Anteroposterior and lateral views of upper extremities and AP views of both hands and wrists.

- Normal epiphysis, U or V shaped physis and flaring of metaphysis are seen in Achondroplasia and Pseudoachondroplasia.

- Platyspondyly is seen in Pseudoachondroplasia, SED and Kniest dysplasia.

- Delayed ossification of epiphysis, fragmentation of epiphysis, irregular and flattened epiphysis are seen in Pseudoachondroplasia**,** SED, Multiple epiphyseal dysplasia (often associated with AVN of the femoral head) and in Kniest dysplasia. CT & MRI are required to assess the spine, and sometimes to assess the morphology of the joints.

AIMS OF TREATMENT

The main aim of the treatment is to optimize the function by correcting the deformities, alleviating pain and preventing future limitations. Restoration of height is no longer considered as the main aim of treatment, as this involves a number of childhood years, loss of schooling and connection with peers**.**

OTHER CONSULTATIONS

The other consultations required are neurosurgery, ENT, cardiology, rehabilitation and genetics.

CURRENT CONCEPTS OF PAEDIATRIC OSTEOPOROSIS: DIAGNOSIS AND TREATMENT

Elizabeth A. Szalay MD

INTRODUCTION

Osteoporosis is increasingly recognized as a factor in a multitude of childhood conditions, yet it is poorly understood and frequently overlooked in children. *"Osteoporosis is a disease characterized by low bone mass, microarchitectural deterioration of bone tissue leading to enhanced bone fragility, and a consequent increase in fracture risk."*[1]

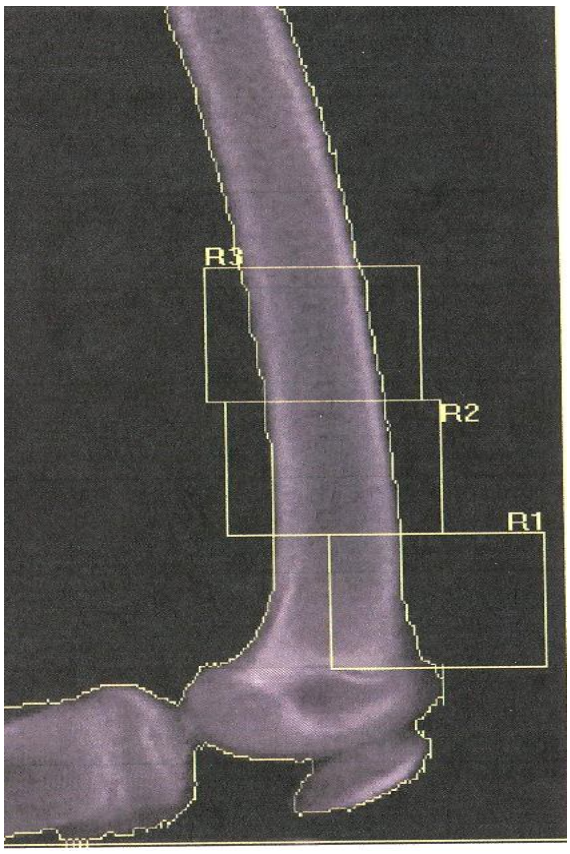
Dual energy x-ray absorptiometry (DXA) is the most widely used tool for assessment of bone mineral density (BMD) and is considered the preferred method because of its precision, reproducibility, speed, and minimal radiation exposure.[i] Use of other methods, such as peripheral quantitative computed tomography and quantitative ultrasound, is limited in children because of scant reference data.[2]

DXA scanning is well adapted to paediatric applications because of its low radiation exposure (3 mrem, as compared, for example, with a lateral lumbar plain film at 70 mrem). The reference databases in children are relatively small (for instance, the Faulkner database[3] consists of 892 scans of the proximal femur and 666 scans of the lumbar spine), but nonetheless they seem to provide clinically relevant information. In children it is a valuable clinical tool, but, with respect to the acquisition and interpretation of the scans, children are not simply "small adults." Unlike the adult osteoporosis population who are commonly healthy older adults, the children most likely to have low bone density for age are likely to have physical challenges such as cerebral palsy or spina bifida that may result in contractures that preclude positioning for spine or hip scans.

Elizabeth A. Szalay, MD,
University of New Mexico,
Carrie Tingley Hospital,
Albuquerque, New Mexico.

The International Society for Clinical Densitometry (ISCD) recommends that scanning of the total body and the spine be done in children [1], but this is impractical in many ill children because of contractures or other physical limitations. Whole body scans are difficult to interpret and are often subject to artefacts. For instance, orthopaedic hardware anywhere in the body renders the whole body scan inaccurate.[4]

The distal lateral femoral scan described by Harcke[5]and Henderson[6] has been found to be an invaluable adjunct in the paediatric DXA practice. It can be done in children with significant contractures, and is less threatening to children because they may lie on their side and hug a pillow or a toy. Many mentally challenged children who



Lateral distal femoral DEXA scan

cannot tolerate an examination in the supine position can often tolerate this examination. An additional advantage is that in quadriparetic children, this is often the site of pathologic fracture and thus a logical choice for bone density measurement. Many centres consider the lateral distal femoral (LDF) scan to be highly reproducible, and use it on most children.

Regardless of the regions of interest to be scanned, special paediatric software is needed to do DXA scans in children, and the individual who interprets the scan must be cognizant of the multiple factors that can impact paediatric bone density.

In adult patients, "osteoporosis" is defined as bone mineral density (BMD), as demonstrated by dual photon X-ray absorptiometry (DXA) scanning, of less than 2.5 SD below the young adult mean, and "osteopenia" is less than 1.0 SD below the young adult mean (the "T-score"). Children do not achieve their adult bone density until the third decade. Hence the use of "T-score" is inappropriate for children. Indeed, according to the International Society for Clinical Densitometry (ISCD), there is no densitometric definition of osteoporosis in children because, unlike in adults, it is not known at what point a significant increase in fracture risk occurs. To label a child as having "osteoporosis," she/he must have exhibited evidence of an insufficiency fracture, which is a fracture sustained as a result of minimal trauma.[1]

Similarly, the term "osteopenia" is not recommended in children, again because fracture thresholds have not been established in children for specific Z-scores.[1] The term recommended by the ISCD is "low bone density for age," and is defined as a bone mineral density that lies less than 2 standard deviations (SD) below the age-and-sex matched mean (the "Z-score").

INSUFFICIENCY FRACTURES

An "insufficiency fracture" is defined as a fracture that occurs with minimal trauma. This may mean a fall from a standing height, spinal compression fractures from jumping or a hard step down, or, in the case of physically challenged children such as those with non-ambulatory cerebral palsy, fractures may occur during simple dressing or donning of orthotics. In children with neuromuscular disorders, insufficiency fractures are commonly seen in the distal femur or proximal tibia.[7,8]

Children and adolescents who undergo hip or pelvic surgery may be immobilized in body casts during the post-operative period or they may be kept non-weight-bearing for prolonged periods of time, and may experience post operative fracture.[9] Children, being metabolically active because of skeletal growth, exhibit an impressive rate of change with respect to bone mineral density. A 2008 study[10] demonstrated in 15 children a loss between 5% and 34% of preoperative bone mineral density (BMD) during the 4-11 weeks following orthopaedic surgical procedures, with an average loss of 16%. The cancellous region of the distal femur, frequently the site of post operative fractures, was seen to drop >1 SD in Z-score. Follow-up DXA scans have shown that it takes several months to replace the bone density even-though it is lost so quickly.

Finally, inexplicable pain in children who cannot communicate verbally has been correlated in some instances with "bone pain" that has responded to bisphosphonate therapy. Bone density of the distal femur has been correlated with fracture risk in non-ambulatory children.[11]

TREATMENT OPTIONS

Because it is not clearly demonstrated at what point (i.e. at what Z-score, which describes how far above or below the mean of age-and-sex-matched controls) a child experiences increased fracture risk, treatment for "low bone density for age" generally consists of supplementation with Vitamin D and calcium.

Children with demonstrated osteoporosis (low bone density plus the occurrence of an insufficiency fracture) are treated with Vitamin D and calcium supplementation as needed, and these patients may be considered for bisphosphonate therapy. Such patients include children with non-ambulatory

cerebral palsy, seizure disorder, spina bifida and muscular dystrophy.

Calcium

Daily calcium requirement varies with age, from about 300 mg/day at age 1 year, 800-1000 mg/day at age 10 and 1200-1500 mg/day for teens and adults. Calcium may be obtained from either dietary sources (240 cc of milk contains about 300 mg of calcium) or from supplementation.

Calcium supplements come in many forms, pills or tablets most commonly, and also in liquid and chewable forms. Calcium carbonate is the most economical form; calcium citrate should be used in individuals lacking gastric acidity, individuals on gastric acid blocking medications, and in those who have had calcium renal stones. Large calcium tablets may be so hard as to not dissolve in the stomach. Bioavailability can be assessed by dropping the tablet into a small glass of vinegar—rapid dissolution in vinegar indicating that the preparation will dissolve in the stomach. Calcium supplements should be taken in divided doses during the day, as intestinal absorption of more than 500 mg per dose is poor.

Vitamin D

Vitamin D is a fat-soluble "vitamin" that is essential for normal calcium metabolism. Technically, it is not really a vitamin, the definition of which being a substance needed in small amounts by the body which the body cannot make.[11] Unlike other vitamins, Vitamin D can be synthesized by humans in the skin upon exposure to ultraviolet B radiation from sunlight, or it can be obtained from diet. "Adequate intake levels" (AI) for Vitamin D were established in 1997 by the US Food and Nutrition Board of the Institute of Medicine[12] and were only recently doubled to 400 International Units (IU)/day for infants and children, a level found to prevent rickets. However this is not necessarily a daily intake sufficient for good bone health.

Most paediatricians assume that children receive adequate amounts of Vitamin D from sunlight. Approximately 15 minutes of sunlight exposure of face, head, and hands is thought to provide adequate amounts of sunlight for skin conversion, while a day at the beach without sunscreen may produce 10,000 IU. Vitamin D is stored in body fat "for a rainy day". Receiving adequate sun exposure is problematic in extremes of latitude, such as Boston or north in the winter, in persons who practice extreme clothing coverage for religious or other reasons, or for those who are of very dark complexion. It can also be seen in "healthy" children who do not play outdoors.[14] Physically challenged children, like those in wheelchairs, who cannot voluntarily run or play in the sun, are unlikely to receive adequate solar exposure.

It is almost impossible to reach adequate serum levels of Vitamin D from diet alone. The only foods that contain appreciable levels of Vitamin D are milk, because it is fortified in the US, and in oily fish. Other foods that are fortified are fortified at very low levels. Children's multivitamins contain 400 IU of Vitamin D or less, in keeping with government recommendations to prevent rickets.

Children with low bone density for age or osteoporosis should be evaluated for Vitamin D status by assessing the serum 25 OH Vitamin D levels.

Supplementation of Vitamin D can be done with daily doses (often requiring 1000-5000 IU/day depending on patient size), or larger doses can be given weekly or monthly. 25 OH D levels should be reassessed in 6-8 weeks. Vitamin D is a fat-soluble vitamin, so overdosage is possible, but toxicity in adults has not been demonstrated in doses <10,000 IU/day.

Bisphosphonate therapy

Pamidronate is a second-generation intravenous bisphosphonate that inhibits calcium absorption from the bone by affecting both the osteoclast and the osteoblast. It has been used extensively in children with osteogenesis imperfecta, juvenile idiopathic osteoporosis, and also to a lesser degree in children with fibrous dysplasia, cerebral palsy and other childhood illnesses resulting in low bone

density. In all of these conditions it has been found to have a good safety profile. In animal studies, at doses recommended for the treatment of hypercalcemia, pamidronate inhibits bone resorption apparently without inhibiting bone formation and mineralization. Henderson et al. in a pilot study using pamidronate in children with cerebral palsy, who had very low bone density, demonstrated an average 89% improvement in BMD in 18 months.

Pamidronate is approved in many countries for treatment of osteoporosis, cancerous bone disease in patients with multiple myeloma or breast cancer and for the treatment Paget's disease of bone. It has not been approved by the FDA for use in children, although studies are proceeding regarding its use in osteogenesis imperfecta.

Pamidronate has been extensively studied abroad and has become the accepted treatment both abroad and in the United States for children with osteogenesis imperfecta. The safety and efficacy demonstrated in osteogenesis imperfecta has resulted in expanded clinical indications, which now include idiopathic osteoporosis and osteopenia of chronic disease such as cerebral palsy, muscular dystrophy and other childhood disorders such as chemotherapy, glucocorticoid therapy or anti-seizure medications. The most commonly used regimen for treatment of osteogenesis imperfecta is 9 mg/kg/year, given IV in 4-6 doses.

In all published studies, side effects of pamidronate have been minimal with the doses used in most clinical settings. Recent reports that have looked specifically for the more alarming side effects such as osteonecrosis of the jaw, have reported no such cases in children or adolescents, even in patients treated for as long as 15 years. Brumsen et al. (Medicine, 1997; 76(4):266-283), reviewing the literature and their own 12 patients treated continuously with pamidronate for up to 8 years, stated that "the administration of nitrogen containing bisphosphonates, pamidronate and olpadronate to children with severe osteoporosis was devoid of any adverse effect in the growing skeleton." Indeed, the indication in osteogenesis imperfecta has been expanded to infants, and administration is begun at three months of age (Astram et al. Arch Dis Child 2007; 92: 332-338.).

Pubmed yields more than 100 publications regarding the use of pamidronate and other bisphosphonate medications in the treatment of childhood conditions resulting in low bone density. Reported adverse effects are few, even with prolonged treatment (Vallo et al, Acta Paediatrica, 2006, 95; 332-339).

The 4th International Conference on Children's Bone Health (Montreal, June 2007), offered 8 presentations on the use of pamidronate in children, including indications of osteogenesis imperfecta, juvenile idiopathic osteoporosis, cerebral palsy, chronic multifocal osteoporosis, Pompe's disease, avascular necrosis following glucocorticoid therapy, and Duchene muscular dystrophy. Safety and efficacy were confirmed, side effects were few, and two papers confirmed that avascular necrosis of the jaw has not been reported in children receiving bisphosphonate therapy. (All abstracts published in Bone 2007: 40, S30-77).

Children treated with pamidronate sometimes experience a slight increase in body temperature accompanied by back and limb pain. This is controlled with acetaminophen. Other side effects that are rare (<1%) but possible are eye irritation and excessive weight gain in adolescents.

There are no established doses for bisphosphonates. Protocols have been developed to give 1-3 mg/kg pamidronate, administered IV over four hours, every one to three months.[15] Total recommended duration of therapy has not been established.

Alendronate and other oral bisphosphonates have also been used. Empiric dose for alendronate is 1 mg/kg per week up to an adult dosage of 70mg/week Response to bisphosphonates should be monitored by DXA scans, ideally once yearly.

CONCLUSIONS

Low bone density and osteoporosis (demonstrated by fragility fracture) should be diagnostic considerations in children when appropriate. These entities may be quantified by DXA scan and by determination of serum 25 OH Vitamin D levels, and should be treated.

"Low bone density for age" (DXA Z-score <2 SD below age and sex matched mean) may be treated with calcium and Vitamin D. Once osteoporosis is established by fracture, bisphosphonate therapy has been well tolerated in children.

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CROUCH GAIT IN CEREBRAL PALSY

Dhiren Ganjwala

INTRODUCTION

Crouch gait is a term which is used to describe a gait pattern characterised by increased knee flexion in the stance phase of the gait cycle. It suggests the failure of the mechanisms responsible to maintain the body in an upright position. The three muscle groups known as the *antigravity muscles* are the hip extensors, the knee extensors and the ankle plantar flexors. These muscles are primarily responsible for keeping the body in the upright position. Inability of one or more of these muscles to function adequately results in a crouch gait and is observed as body collapsing into a flexion posture.

To differentiate crouch gait from other knee flexion gait patterns, three parameters which are seen during the late stance phase are used to define crouch gait -

1. Increased ankle dorsiflexion
2. Increased knee flexion and
3. Diminished hip extension

Observational or video gait analysis is adequate to identify a severe crouch gait. However a borderline abnormality may require an instrumented gait analysis. The exact incidence of this gait abnormality in a diplegic population is not known. However gait evolution studies suggest an increase in the incidence of crouch gait during adolescent years. This article describes the causes, ill effects, prevention and management of crouch gait.

RELEVANT BIOMECHANICS

The following three muscle groups maintain erect posture. (Fig 1)

1. Hip extensors (*gluteus maximus and hamstrings*) generate about 20-30% of force necessary to maintain erect posture. This group is mainly active during the initial third of the stance phase.
2. The knee extensors (vasti) generate about 20-30% of force. This group is also active during the initial third of the stance phase.
3. The ankle plantar flexors (soleus) generates about 40-50% of the force required to maintain an erect stance. The soleus pulls the tibia backwards and stabilizes it in relation to the foot. Ground reaction force then carries out knee extension. This is known as the "ankle plantar flexion/knee extension couple" (Fig 1). This mechanism maintains the erect posture during the later part of the stance phase.[1]

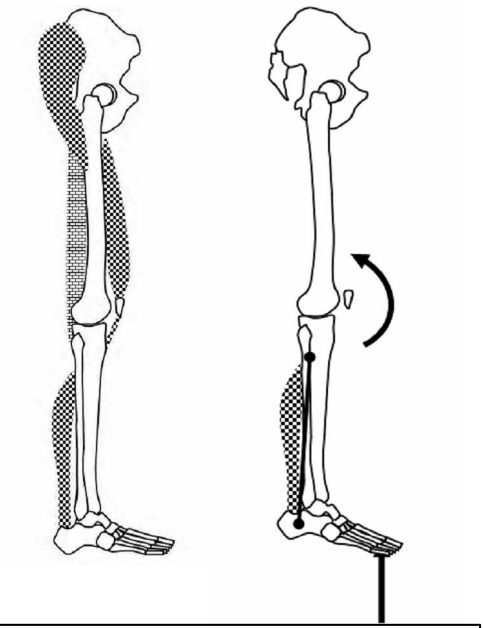

Fig 1: Left showing antigravity muscles maintaining limb position in stance. Right showing action of soleus pulling tibia backwards and plantarflexing ankle

CAUSATIVE FACTORS

Lever arm dysfunction

This is based on the principle of levers for effective functioning. When the lever arm is short, muscles do not produce adequate power. Bone and joint pathology like torsion deformity of long bones or planovalgus deformity of the foot reduce the lever arm. This group of etiological factors are termed

Dhiren Ganjwala, M.S. (Ortho)
Ganjwala Orthopaedic Hospital,
302 Anshi Avenue, Opp.Mrudul Park, Polytechnic,
Ahmedabad - 380 015
Gujarat, India
Email: ganjwala@gmail.com

"lever arm dysfunction (LAD)". As the child develops planovalgus foot deformity, the foot becomes unstable and is not able to provide a rigid lever. This is similar to lifting a load with a rubber crowbar. [2,3] (Fig 2)

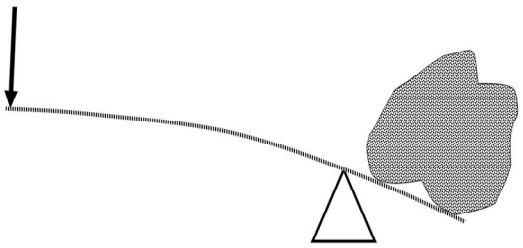

Fig 2

Joint deformity

Flexion deformity at the hip/knee or dorsiflexion at the ankle cause a crouch gait. These joint deformities can be fixed or dynamic. Knee flexion deformity is one of the common findings in crouch gait. There is no compensatory mechanism other than to crouch in the presence of a knee flexion deformity.

Muscle Overstretching

The Quadriceps mechanism stretches due to prolonged ambulation with knee flexion. Subsequently, even with correction of the knee flexion deformity by operative or non-operative means the quadriceps remains elongated unable to carry out the last few degrees of active knee extension producing an extensor lag. Similarly the elongated soleus is unable to restrain the forward movement of the tibia during the second rocker (Fig 3). This results in excessive dorsiflexion at the ankle resulting in a compensatory increase in the flexion at the hip and knee.

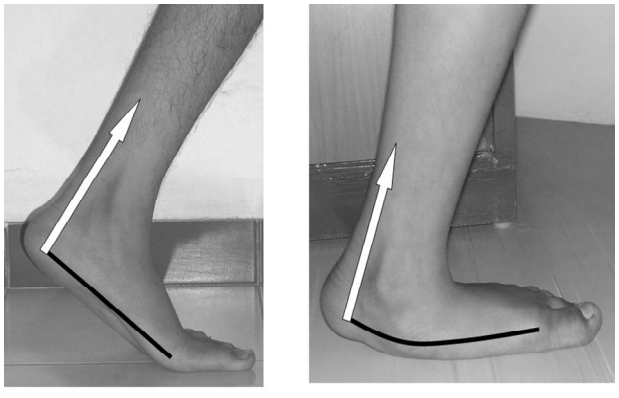

Fig 3: Stable foot providing rigid lever arm and planovalgus foot mimicking flexible lever arm

Muscle weakness

Weakness is one the primary impairments in cerebral palsy worsened by lack of physical activity, which is very common in diplegic children. Few reasons specific for soleus weakness are:

- Tendoachilles lengthening when only the gastrocnemius is short results in an undesirable lengthening of soleus resulting in weakness.
- Prolonged use of an Ankle Foot Orthosis.
- Repeated use of Botulinum injection.

The vasti and hip extensors have to work more to compensate for a weak soleus, so as to maintain the erect posture. When more power is required for hip extension, hamstrings join the gluteus maximus. In erect posture with hip and knee extended the lever arm for hamstrings is longer at the hip than at the knee. But with hip and knee in flexed position, as seen in crouch, the lever arm at knee becomes longer compared to that at the hip. Hence, hamstrings now function more effectively as knee flexors than as hip extensors (Fig 4).

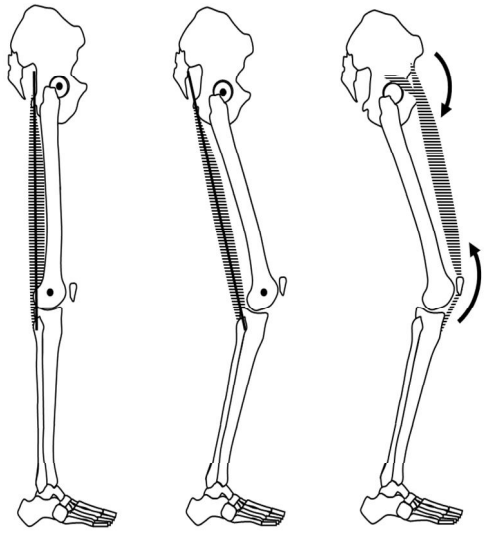

Fig 4: Effect of hip and knee flexion on lever arm as seen in crouch. With increasing knee flexion, lever arm of hamstrings as knee flexor increases and with increasing hip flexion lever arm for rectus as hip flexor increases.

To counter the hamstrings, more power is required to extend the knee for which the rectus starts to reinforce the vasti. But rectus femoris is both a knee extensor and a hip flexor. So when it is acting to stabilize the knee, it also causes flexion at hip. This further accentuates the crouch deformity.

Adolescent growth spurt

During adolescent growth spurt, height and weight of the child increases more than the strength of muscles. Prior to adolescent growth spurt, muscles which were strong enough to maintain erect posture, fail to maintain erect posture after adolescent growth spurt.

EFFECTS

Most of the ill effects of crouch gait are seen at the knee. When one walks with knee in flexion, one requires more quadriceps power. Walking with 30 degrees of knee flexion requires the quadriceps muscle to work at around 50% of its maximum power.[4] The cyclic demand for extra power promotes fatigue at lesser distances. Another reason for early fatigue is the necessity of the quadriceps to stabilize the knee in the entire stance phase. Designed to be active only in the initial third of the stance phase, persistent knee flexion throughout the stance tends to buckle it.

Standing with more than 30 degrees of knee flexion puts excessive tensile force on the patellar tendon which results in an elongated patellar tendon, and consequently results in either a patella alta, or a stress fracture at inferior pole of patella or tibial tubercle.[5] In one study, 21% of the immature, ambulatory children with spastic cerebral palsy had such radiological abnormalities.[6]

Walking with knee flexion also increases loading of the patello-femoral joint. Needless to say, this results in pain and an increased risk of later date patello-femoral arthritis.

The above mentioned effects work in tandem leading to gait deterioration, increased dependence on support and sometimes even the need of a wheelchair to aid ambulation.

PREVENTION

Crouch gait reduces functional ability of the child. An aggressive effort is the key for controlling crouch gait. Strengthening exercises, mainly for antigravity muscles, should be encouraged from early age. In the diplegic child, bi-articular muscles are more involved than uni-articular muscles. Commonly they have a shortened gastrocnemius and a normal length soleus. Tendoachilles surgery in such a case will lead to unnecessary lengthening of soleus and thus weakens it. Strayer's or Baumann's procedure which can selectively lengthen gastrocnemius is preferred to avoid iatrogenic weakening of soleus.

Lever arm deformities like planovalgus foot can be prevented or can be controlled well by effective use of orthosis. If the child has a tendency to develop a crouch then ground reaction orthosis should be used to support the upper leg from front so as to avoid forward displacement of tibia (Fig 5).

Knee flexion deformity should be prevented by use of stretching splints. Children have tendency to gain weight during adolescent years. Muscles which were able to maintain upright posture when child was young may fail to maintain upright posture with weight gain. Hence weight gain should be monitored and avoided.

TREATMENT

Crouch can be a serious problem for a child. It cripples the child and significantly decreases his capacity to walk. All factors causing crouch should be identified and managed. Following factors need surgical treatment.

- External tibial torsion can be managed by derotation osteotomy at distal tibia. Similarly femoral torsion can be corrected by derotation osteotomy.

- Planovalgus foot is managed by lateral column lengthening.

- If there is instability at midfoot, fusion of calcaneo-cuboid joint is required.

- If hindfoot valgus is not completely corrected with lateral column lengthening then medial displacement osteotomy of calcaneus is carried out.

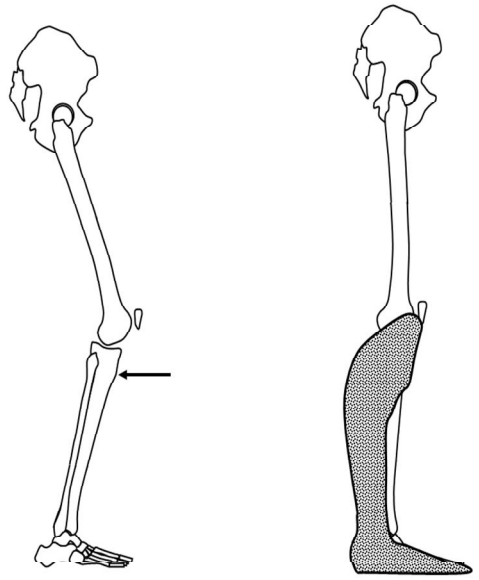

Fig 5: Ground reaction orthosis used to prevent crouch gait

If the child is walking with crouch gait for a long time the child may have developed flexion deformity at the knee. Knee flexion deformity is corrected by capsulotomy or temporary anterior epiphysiodesis by staple or 8 plate.[7] When limited growth potential is remaining and deformity is more than 20° supracondylar osteotomy is preferred. [8]
Elongated patellar tendon is an important cause of failed treatment and that needs to be addressed. Patellar tendon plication or distal transfer of tibial tuberosity can effectively take care of over stretched muscles. When the child is young and skeletally immature one can correct other factors and use ground reaction orthosis for few months so that the longitudinal growth can reduce the slack of patellar tendon.[9]

Weak muscles: Psoas, hamstrings and gastrocnemius are lengthened if they are short on clinical examination. In addition to surgical treatment, strengthening exercises of all anti gravity muscles should be carried out.

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FEMORO-ACETABULAR IMPINGEMENT: RADIOGRAPHIC EVALUATION

Smitha E Mathew and Vrisha Madhuri

INTRODUCTION

Femoroacetabular impingement (FAI) is a recently described condition in which an abnormally shaped proximal part of the femur or an excessive acetabular coverage causes abutment between the femoral head-neck junction and the acetabulum. It has been recognized as a cause of early osteoarthritis in young adults.

Two mechanisms of impingement are:

1) **Cam-type impingement**: due to a *non spherical* femoral head.
2) **Pincer-type impingement**: caused by *excess acetabular coverage*.

CAM IMPINGEMENT

Radiographic views

i) Cross table lateral view –

This view is obtained by placing the affected hip in 15° of internal rotation and flexing the opposite hip (Fig 1). The x-ray beam is parallel to the table from opposite side, oriented at 45° to the symptomatic limb, and the crosshairs directed at the center of the femoral head.

ii) 45° Dunn view –

This is the most sensitive radiographic view. With the hip flexed 45°, abducted 20° and in neutral rotation, the beam is directed from above to a point midway between the ASIS and the pubic symphysis (Fig 1).

iii) 90° Dunn view –

This view is performed in a similar fashion, with the only exception being that the hip is flexed to 90°. It is not done routinely due to difficulty in maintaining this position.

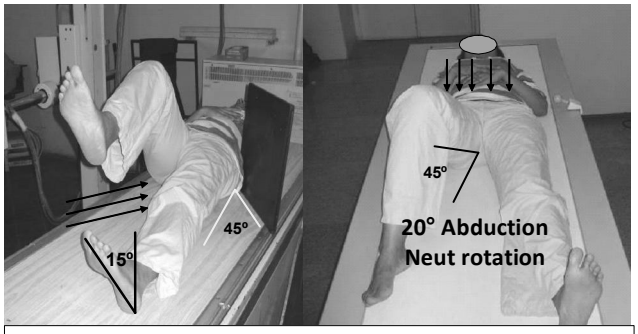

Fig 1: Position for Cross-table lateral view (Left) and 45° Dunns view (Right)

iv) Frog-leg Lateral view –

With the knee flexed 30°–40°, hip abducted to 45° and the heel resting against the medial aspect of the contralateral knee, the beam is directed from above to a point midway between the ASIS and the pubic symphysis (Fig 2).

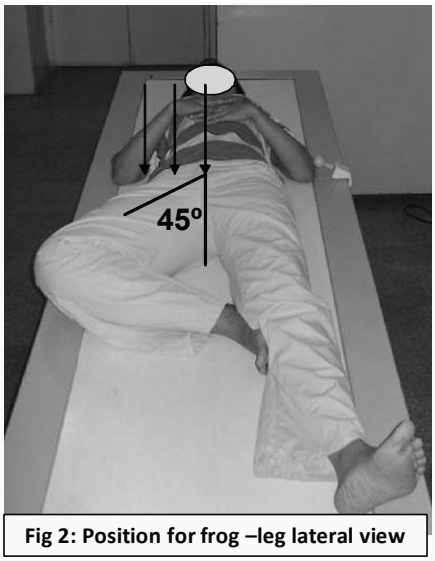

Fig 2: Position for frog –leg lateral view

Interpretation

i) α angle -

Frog-leg lateral view is used to assess this angle. The first line is drawn connecting the center of the femoral head and the center of the femoral neck. The second line is drawn from the center of the femoral head to a point on the anterolateral head-

Smitha E Mathew, Tutor
Vrisha Madhuri, Prof & Head
Paediatric Orthopaedics Unit
Christian Medical College, Vellore – 632 004
Tamilnadu, India
Email: madhuriwalter@cmcvellore.ac.in

neck junction where the radius of the femoral head begins to increase beyond the radius found more centrally in the acetabulum where the head is more spherical (i.e., where a prominence or bump starts). The intersection of these two lines forms the *alpha angle* (Fig 3). An angle >50.5° is considered abnormal.

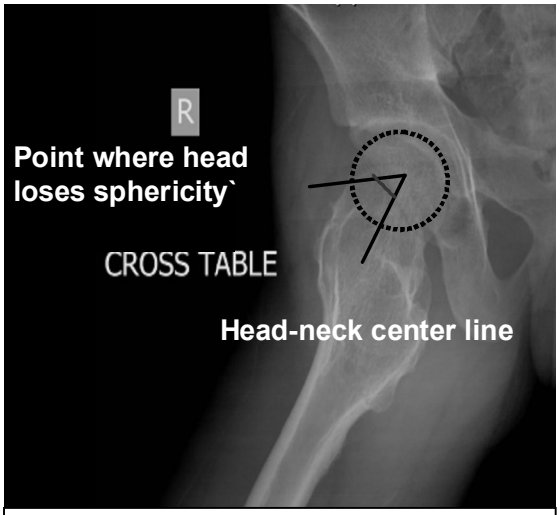

Fig 3: Radiograph demonstrating α angle

ii) Anterior offset -

This is the distance between two parallel lines, one adjacent to the anterior aspect of the neck and the other touching the anterior most part of the femoral head, both being parallel to the femoral neck axis (AB) (Fig 4). The value in asymptomatic hips is 11.6 ± 0.7 mm; Hips with cam impingement will have a value of 7.2 ± 0.7 mm.

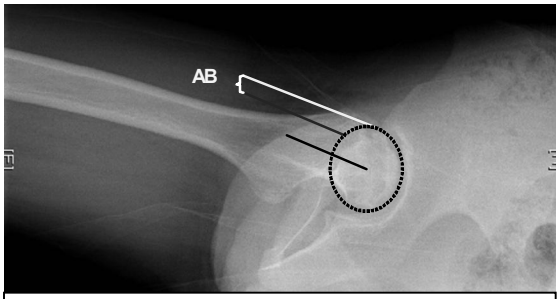

Fig 4: Lateral hip radiograph showing method of assessing anterior offset

iii) The head-neck offset ratio -

The ratio is determined by three parallel lines drawn similarly as shown in Fig 4. Line 1 is drawn through the center of the long axis of the femoral neck; Line 2 drawn through the anterior most aspect of the femoral neck ; Line 3 drawn through the anterior most aspect of the femoral head.

The head-neck offset ratio =

$$\frac{\text{Distance between lines 2 and 3}}{\text{Diameter of the femoral head}}$$

If the ratio is <0.17, a cam deformity is likely present.

iv) Head sphericity-

With use of an AP pelvic, a 45° or 90° Dunn, a frog-leg lateral, and/or a cross-table lateral view, the femoral head can be classified as either *spherical or aspherical*.

A Mose template (concentric circles) can also be used as a reference. The femoral epiphysis is termed aspherical if the asymmetry is > 2 mm and it is termed spherical if asymmetry is < 2mm.

PINCER IMPINGEMENT

Radiographic views

i) False profile View of Lequesne and de Sèze -

This view is done with patient in standing position. The affected hip is placed against the cassette and the pelvis rotated 65° in relation to the bucky wall stand. The foot on the same side as the affected hip should be parallel to the cassette. Beam is directed from the front to a point midway between the ASIS and the pubic symphysis (Fig 5).

Fig. 5: Clinical picture demonstrating patient's position for False Profile view

ii) Anteroposterior pelvic view -
Hips are placed in 15° internal rotation to compensate for femoral anteversion and to provide better visualization of the contour of the lateral femoral head–neck junction.

Beam is centered on the point midway between the superior border of the pubic symphysis and a line drawn connecting the ASIS.

Interpretation
i) "Crossover" sign -
This sign is seen in a retroverted acetabulum. Acetabular retroversion is defined as anterior wall (AW) being more lateral than posterior wall (PW), whereas in a normal hip anterior wall lies more medially (Fig 6).

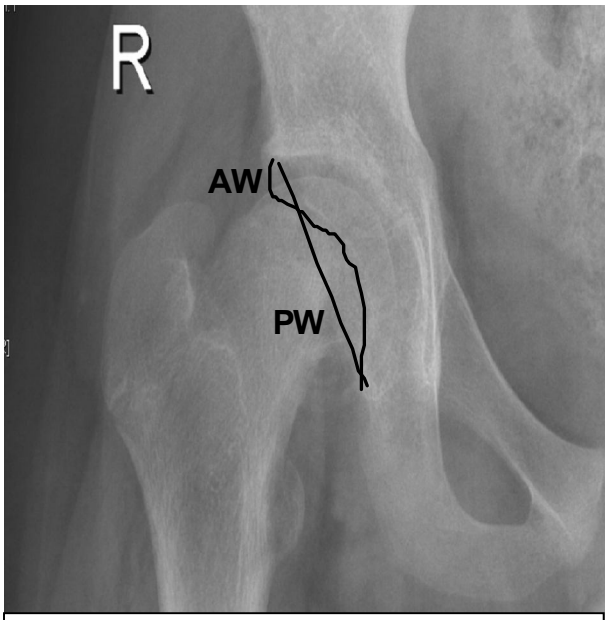

Fig. 6: Right hip showing "Crossover" Sign

ii) Acetabular Depth -
It is assessed in anteroposterior pelvic radiograph. The relationship of the floor of the acetabular fossa and the femoral head should be evaluated relative to the ilioischial line.

a) Coxa profunda
In AP pelvic radiograph the medial wall of the acetabulum lies on or medial to the ilioischial line.

b) Protrusio acetabuli
Represents the more severe form of coxa profunda, and is diagnosed when the femoral head crosses the ilioischial line.

iii**) Posterior wall sign -**
This sign is considered to be present when the center of the femoral head is lying lateral to the posterior wall (Fig 7).

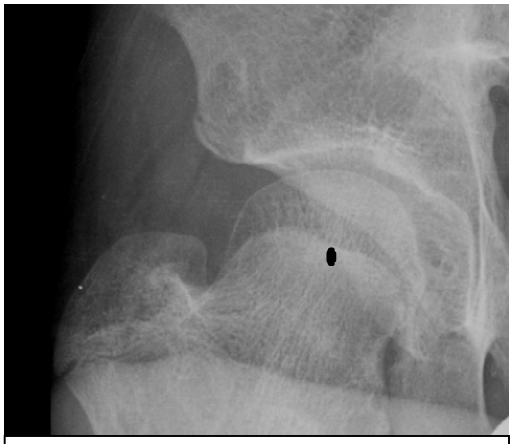

Fig 7: Posterior wall sign

iv) Lateral center-edge (CE) angle, or CE angle of Wiberg -
This angle is assessed in AP pelvic radiograph and it indicates the superolateral coverage of the femoral head by the acetabulum. The angle formed by a line drawn from the center of the femoral head to the

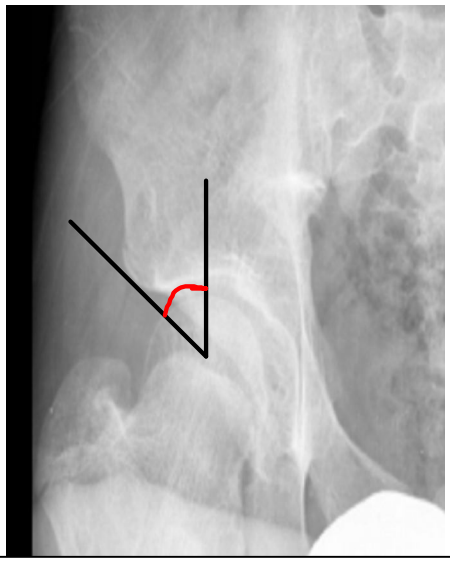

Fig.8: Lateral CE angle assessed in AP view

outer edge of the acetabular roof, and a vertical line drawn through the center of the femoral head (Fig.8). An angle >39° is an indicator of acetabular over-coverage.

v) Anterior CE Angle of Lequesne -
Anterior coverage of the femoral head is assessed in a *false profile radiograph.* It is also used to assess the postero-inferior part of the hip joint to detect the so-called contrecoup lesions in pincer impingement.

This angle is formed by a vertical line through the centre of the femoral head and a second line through the centre of the hip and the foremost aspect of the acetabulum (Fig 9). The normal angle is 26°- 25° and is increased in FAI.

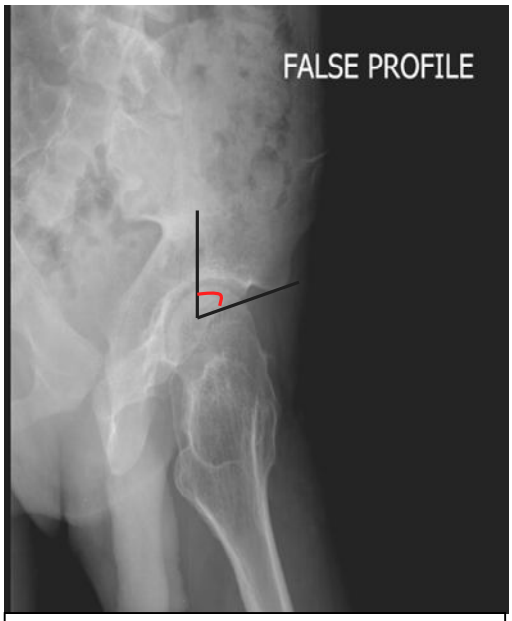

Fig.9: Anterior CE angle assessed in FP view

vi) Acetabular index:
Angle formed by a horizontal line and a line connecting the medial point of the sclerotic zone with the lateral edge of the acetabulum.

In hips with coxa profunda or protrusio acetabuli, the acetabular index (also called "acetabular roof angle") is typically 0° or even negative.

vii) Pitt's Pit -
Fibrocystic changes at the femoral head-neck junction are termed as Pitt's pit (Fig 10). Though they are often visible on plain x-rays, they are more easily seen on CT or MRI scans. They are reported to be 91% specific and to have a positive predictive value of 71% for FAI.

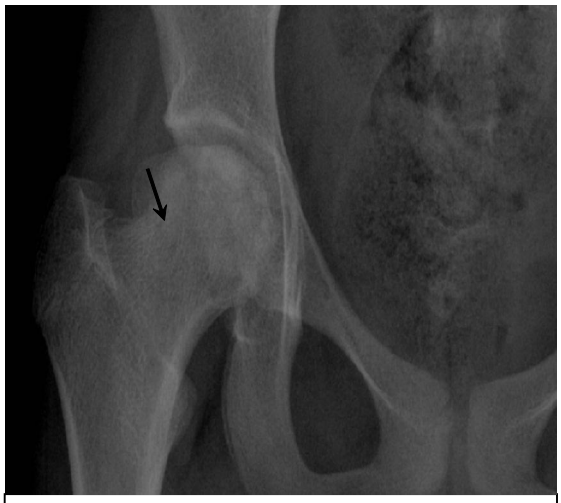

Fig.10: AP view of hip showing degenerative changes in femoral head

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ANTIBIOTIC DURATION IN PEDIATRIC CHRONIC OSTEOMYELITIS: WHAT IS THE EVIDENCE?

Abhay Gahukamble

INTRODUCTION

Chronic osteomyelitis in children is a debilitating disease in the developing world which places strain on the already burdened health system. The aim of this review is to assess the evidence available to guide management especially with respect to antibiotic therapy and outline issues requiring further research.

Broadly the issues to be addressed are the role of antibiotics, route of administration, duration of therapy and briefly the role of debridement and evaluation of its adequacy in the clinical setting and the outcome measures used to evaluate success of a regime will be considered.

SEARCH STRATEGY

The Cochrane library Issue 3, 2009; Pubmed and Proquest medical library were searched for randomized and quasi-randomized trials with regards to antibiotic usage using the search term "chronic osteomyelitis" and Mesh Search ("Osteomyelitis" AND "chronic") with limits "All child 0-18 years" in Pubmed. The reference lists of relevant articles were hand searched.

RESULTS

The search revealed 47 clinical trials inclusive of 8 randomized control trials. Of these, five trials[1-4] were considered to be relevant. The trial populations were predominantly adults with a minority of children. One of these articles comparing Corydalis saxicola bunting liquor combined with antibiotics was in Chinese[5] and could not be accessed, as was a clinical trial of cloxacillin and tetracycline in Ugandan children with staphylococcal osteomyelitis[6] published in 1973.

Giamarellou et al[1] evaluated pefloxacin versus ceftazidime in a series of gram negative infections comprising 15 chronic osteomyelitis. This prospective open randomized trial concluded that pefloxacin is as effective as ceftazidime in moderate to severe gram negative infections.

Patzakis et al[2] evaluated 35 patients with chronic osteomyelitis of which 12 were randomized to the septopal group and five to the control group. However since there was just one patient 18 years of age, the applicability of this study to pediatric chronic osteomyelitis is questionable.

Meissner et al[3] evaluated treatment of chronic osteomyelitis with two doses of fosfomycin in 60 patients and reported results at a mean of 37 months. Norden et al[4] compared parenteral nafcillin versus parenteral nafcillin with oral rifampicin and noted that 70% of patients with the latter regime experienced apparent cure.

DISCUSSION

The vast majority of management regimen of chronic osteomyelitis in children is based on Level III and IV evidence. No clinical trials dealing with children exclusively were found in this area. Children with chronic osteomyelitis will have a different natural history as compared to their adult counterparts and hence it would not be prudent to extrapolate results obtained from clinical trials in adults with chronic osteomyelitis to children. Large case series and non-controlled non-randomized prospective trials have an important contribution in this current scenario where adequate high quality evidence is not available. They help in elucidating the natural history, side effects and success/failures of their respective regimen. They are also indispensible in hypothesis generation to allow planning of ethical study designs. However the possibility of bias as well as differences in morbidity and mortality in different centers do not allow any comparison between them. We were unable to find adequate evidence to recommend guidelines regarding the duration and route of antibiotic therapy. The empirical six weeks based on the observation that bone requires that amount of time to revascularize may be followed till availability of evidence to the contrary. Swiontkowski et al[7] noted no difference in long term parenteral therapy as

Abhay Gahukamble
Assistant Professor,
Paediatric Orthopaedics Unit,
Christian Medical College, Vellore – 632 004
Tamilnadu, India.
Email: abhaygahukamble@gmail.com

compared to step down oral antibiotics in adults with chronic osteomyelitis.

The drawbacks with this review are that the search was restricted to Pubmed and the Proquest library. Hence there is a possibility of missing trials indexed in EMBASE and LILACS as well as regional databases. There are various issues with conducting clinical trials in chronic osteomyelitis.

Role of debridement and assessment of its adequacy

Chronic osteomyelitis is characterized by the presence of necrotic bone which forms a barrier to effective antibiotic therapy. Thus debridement has been considered to be of prime importance, though Reinher et al[8] have challenged its necessity in all cases. The adequacy of debridement is difficult to quantify and though Swiontkowski et al[7] have studied and applied laser doppler flowmetry for the same, it is not feasible in most of our clinical settings. Thus there is the issue of patients not being equally susceptible to failure of therapy at baseline in case of non randomized trials. Randomization after debridement should equally distribute the patients eliminating the possibility of bias due to different degrees of debridement if appropriately done.

Role of antibiotics: duration and route of administration

Duration of antibiotic therapy is empirical and to date there is no evidence to show any clear difference between short and long term regimen of sensitive antibiotics. Since deep cultures are available only a few days after surgery the antibiotics used in the interim would need to be based on the local microbiological spectrum. Allocation concealment and blinding to the treatment regimen are issues that need to be dealt with necessitating dummy medications. Retrospective studies assessing recurrence of infection in defaulters to long term therapy will give some insight into the duration of antibiotics to be used in the treatment arms.

Outcome measures in chronic osteomyelitis

Cure as such is not defined in chronic osteomyelitis since recurrence of infection has been described after years of quiescence. Most clinical trials consider quiescence of infection for 1 year as apparent cure and use it as the outcome measure. Infection needs to be defined prior to the onset of the study based on clinical and/or laboratory criteria. Being a dichotomous outcome measure, it would warrant a larger sample size. Thus development of a continuous outcome measure like a validated score for the same will allow smaller more feasible sample sizes but adequately powered studies[9]. Time to recurrence of infection can also be considered as an outcome measure allowing analysis by Kaplan Meier survival analysis.

CONCLUSION

In conclusion, there is a need for significant amount of research in pediatric chronic osteomyelitis. A cheap, practical and reproducible method to assess adequacy of debridement will be beneficial but not absolutely necessary in a randomized trial. Development of a validated score to assess outcome should be considered.

Conducting prospective randomized trials with short term antibiotic regimen would be unethical unless animal studies, retrospective studies in children on short term therapy and those defaulting long term therapy show reasonable evidence to allow the same. Confounders like stage of osteomyelitis, degree of surgery, presence of hardware or any immunocompromised situation need to be addressed while designing prospective studies. Stratification of the important ones is one method to deal with it. Well defined objective outcome measures must be used to decrease bias and blinding the outcome assessor and statistician involved in analysis should be feasible.

Chronic osteomyelitis being relatively common in our country, the onus is on us to generate the evidence to appropriately treat this disease.

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QUIZ

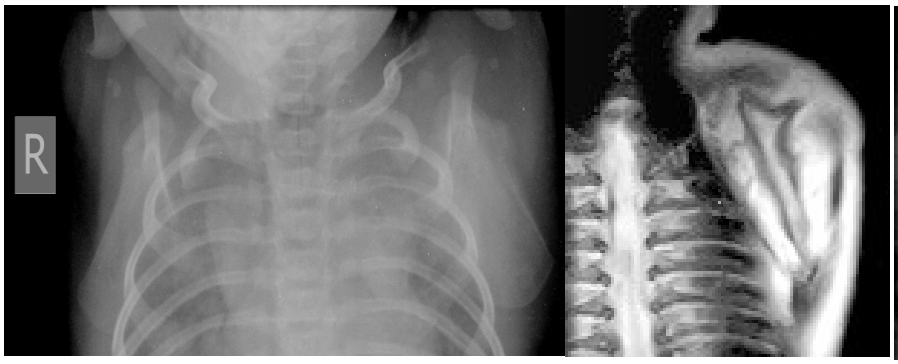
A 4 month old baby was brought by parents with three weeks history of progressive swelling in the region of left scapula, associated with paucity of movements of left upper limb. The child had no associated fever, had not sustained any injury and was apparently normal till the onset of these symptoms. There was no history of involvement of any other site.

On clinical examination a diffuse tender swelling was noted over the left scapular region extending to the left deltoid region. Passive shoulder movements were painful and restricted. There was no regional lymphadenopathy and there were no neurovascular deficits.

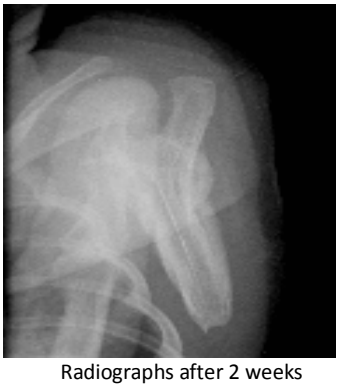
Investigations revealed:

Total WBC Count-16,600 /mm^3
Differential count - N-40,E -8 ,B -0,L -46,M -6
Platelet Count-433,000/mm^3
Hb -9.3 mg/dl
ESR at the end of 60 min -108 mm
Bleeding parameters were within normal range

LDH - 649 U/L
Alkaline Phosphatase -169 U/L
S. Uric acid - 1.9 mg/dl
S.Creatinine -0.4mg/dl
S.Ca -10.4mg/dl, S. Phosphorus -7.7mg/dl



Plain radiograph (left) showing AP view of scapulae and on right MRI: T1W coronal images



Radiographs after 2 weeks

Answer: Next Issue

JOURNAL REVIEW

Harish P, Chirag Bhalvani, Vidyasagar C, Sanjay Chilbule, Balakumar B, Vivek Dutt, Vrisha Madhuri

DIFFICULT-TO-TREAT ORTOLANI-POSITIVE HIP: IMPROVED SUCCESS WITH NEW TREATMENT PROTOCOL

Vineeta T. Swaroop, and Scott J. Mubarak
J Pediatr Orthop 2009; 29 :224-230
Investigation performed at the Rady Children's Hospital, San Diego, CA

This is a retrospective case-control study which assesses the effect of ultrasound in children with dislocated hips. The patients were children under 6 months of age with Ortolani positive hips treated with Pavlik harness. The Intervention was an office based serial USG examination & abduction brace for those hips which were unstable after 3 weeks of harness. The mean follow up of this group was 34 months. The comparison group comprised of children less than 6 months of age who underwent treatment with harness alone, between 1984 and 1997, and the mean follow up period was 28 months.

The primary outcome measure was the success of harness in preventing further treatment. The secondary outcomes that were looked for were the reduction in the rate of surgery and avascular necrosis (AVN) of femoral head.

Results showed that 41 of the 44 hips (93%) in the treatment group were treated successfully. Three hips required further treatment of which 2 underwent operative treatment. None had adverse effects. In the comparison group 44 of the 52 hips (85%) were treated successfully. Eight hips required further treatment of which 5 hips required open reduction. One hip developed AVN of femoral head.

Comments

The prognosis of DDH depends on establishing an early diagnosis and providing appropriate treatment, to achieve and maintain concentric reduction until hip stability is obtained. In the last decade addition of ultrasonography to the management protocol for DDH has influenced the outcome of the treatment in early infancy by allowing confirmation of reduction and assessing stability of the hip in question. It has not only directly improved the rate of reduction with Pavlik harness but also has increased its safety by decreasing the rate of AVN, by confirming concentric reduction and avoiding open reductions for DDH. The results of the above study emphasize the importance of ultrasonography. It is advisable to use ultrasound routinely while managing DDH in infancy.

THE EFFECT OF THE FEMORAL HEAD OSSIFIC NUCLEUS IN THE TREATMENT OF DEVELOPMENTAL DYSPLASIA OF THE HIP: A META-ANALYSIS

Andreas Roposch, Kuldeep K. Stöhr, and Michael Dobson
J Bone Joint Surg Am. 2009; 91: 911-918
Investigation performed at the Great Ormond Street Hospital for Children and the Institute of Child Health, London

Controversy still exists about the role of femoral head ossific nucleus as a risk factor for the development of osteonecrosis of femoral head, following the treatment of developmental dysplasia of hip (DDH). The controversy is regarding the protective influence of ossific nucleus in infancy on the development of avascular necrosis of femoral head, in a child with DDH. There have been reports of studies favouring and refuting the above statement.

In an attempt to resolve the controversy the authors have done a systemic review of Medical literature from 1966 to 2007. They included all the studies in which the outcome measure was osteonecrosis following treatment of DDH in infants or older children. The other inclusion criteria were the studies in which the authors had looked for presence or absence of ossific nucleus in relation to development of osteonecrosis of femoral head. In addition, the other criterion was the studies which looked for presence or absence of ossific nucleus in relation to development of AVN. Only 6 such studies were found and were analyzed.

Paediatric Orthopaedic unit,
Christian Medical College, Vellore - 632 004
Tamilnadu, India
Email: paedortho@cmcvellore.ac.in

A meta-analysis was then performed with the main outcome defined as the development of AVN of the femoral head two years after reduction.

Excluding one case control study, combining the data from all other studies showed no significant effect with the presence or absence of the femoral head ossific nucleus, on the development of AVN grade I-IV of Bucholz and Ogden or Kalamchi & MacEven classification. It occurred in 19% of infants with an ossific nucleus compared with 22% in the group without an ossific nucleus (relative risk = 0.75, 95% confidence interval = 0.46 to 1.21). However if cases of Grade-I radiographic changes of AVN (which some consider as normal) were excluded and grade II or worse were only considered to represent AVN, a significant difference was observed. Only 7% of the infants with ossific nucleus developed AVN, while the incidence was 16% in those without an ossific nucleus (a relative risk of 0.43). This suggests that the development of more severe forms of AVN (grade II or worse) is associated with the absence of the ossific nucleus. Combining data of patients treated with closed reduction from different studies and their analysis showed that the presence of ossific nucleus reduced the probability of developing AVN by 60%. The similar analysis for group treated with open reduction did not show any significant difference. Because the quality of evidence for this finding is moderate, authors concluded that further research is likely to have an important impact on the confidence in these estimates of effect and may well change the estimates.

Comments

Treatment of developmental dysplasia of hip should be started at the earliest. Closed or open reduction need not be delayed waiting for the appearance of the ossific nucleus of femoral head. The issue of presence or absence of ossific nucleus is taken into consideration below the age of one year. This metanalysis included children up to 3 years of age and it is possible that some children in the older age group may have had preexisting AVN thus skewing the outcome for severe forms of AVN. However it is worth studying the influence of presence or absence of ossific nucleus in the development of osteonecrosis. An appropriately designed randomized control trial is required to ascertain the influence of ossific nucleus on the timing of management of DDH.

MEAN 20-YEAR FOLLOWUP OF BERNESE PERIACETABULAR OSTEOTOMY

Simon D. Steppacher, Moritz Tannast, Reinhold Ganz, Klaus A. Siebenrock

Clin Orthop Relat Res. 2008; 466:1633–1644.

The study was conducted in the Department of orthopaedic surgery, Inselspital, University of Bern, Switzerland

Authors are looking at the long-term efficacy and the factors affecting the outcome at an average of 20 years following Bernese Periacetabular osteotomy (PAO). Factors which were looked for were the age at surgery, sex, BMI, Merle d'Aubinge & Postel hip function score and several radiological parameters to assess the osteoarthritis (OA) grade, impingement, coverage and extrusion.

The patients comprised of 63 adolescents and adults (75 hips) who underwent PAO at a mean age of 29 years (13-56 yrs). 31 patients had previous surgeries. Twenty four percent already had advance OA at the time of surgery. The intervention was PAO in all. Concomitant intertrochanteric osteotomy was done in 13 patients (16 hips). The outcome measures were the survivorship of the hips with PAO at mean 20 yrs, maintenance of clinical and radiographic scores, the factors affecting the poor outcome and how the results of hips with PAO fared compared with those of natural course or hips with other osteotomies.

The results showed that the hip score had decreased to the pre-op score. The authors observed no major changes in the radiographic parameters during the 20-year post-operative period except the OA grade which increased from a pre-operative mean score of 0.4 ± 0.6 to a 20-year post-operative mean score of 1.1 ± 1.0.

The authors identified six factors predicting poor outcome: (1) increased age at surgery, (2) lower preoperative Merle d'Aubigne´ and Postel score, (3) positive preoperative anterior impingement test, (4) preoperative limp, (5) preoperative increased OA score (Tonnis), and (6) postoperative increased extrusion index. One was demographic, three were clinical, and two were radiographic factors. A substantial number of these factors were associated

with an already advanced stage of joint degeneration at the time of surgery.

The worse 20-year outcome occurred in patients who were 30 years and older at the time of surgery, patients with preoperative Merle d'Aubigne´ and Postel scores of 14 or less, those with a preoperative OA grade of 2, and those with a postoperative extrusion index of 20% or more.

The authors commented that the surgical technique underwent some minor modifications since the first case in April 1984. The supra-acetabular osteotomy is now placed more cranially from the anterior-inferior to the anterior-superior iliac spine to preserve vascularity of the acetabular fragment and to improve the hold of the Schanz screw for reorientation. An additional arthrotomy is now performed routinely allowing for treatment of labral cysts, observation of range of internal rotation and flexion, and for correction of an aspherical portion of the femoral head-neck offset by osteochondroplasty so as to improve impingement-free ROM. Therefore the clinical outcomes of the current technique can be anticipated to be superior to those in this study.

Comments

This series represents the learning curve of a demanding procedure carried out by the originator. PAO provides an excellent coverage of the femoral head. As the authors suggest, addressing the femoro-acetabular impingement and labral tears at the time of osteotomy would significantly improve the long term results. However careful patient selection and proper surgical technique are important in determining the long-term outcome.

PHYSEAL FRACTURES OF THE DISTAL TIBIA: PREDICTIVE FACTORS OF PREMATURE PHYSEAL CLOSURE AND GROWTH ARREST

Jeffrey T. Leary, Matthew Handling, Marcus Talerico, Lin Yong and J. Andrew Bowe

J Pediatr Orthop 2009;29:356-361.

Investigation performed at the UMDNJ-Robert Wood Johnson University Hospital,
New Brunswic,NJ

The purpose of this study was to determine the incidence of physeal growth arrest and its clinical predictors after distal tibial physeal injuries. The authors retrospectively reviewed 124 patients with distal tibial physeal injuries. They recorded age, sex, mechanism of injury, type of injury according to the Salter-Harris (SH) classification, amount of initial displacement (mm), number of attempts to get reduction, residual displacement (mm) and method of treatment. The mean follow-up was 57 weeks (8-400 weeks) from the time of injury.

Three percent of injuries (4 patients) were SH-I, 32% (40) of the fractures were SH-II, 16% (20) were SH-III, 9% (11) were SH-IV, 19% (23) were triplane fractures and 21% (26) were Tillaux fractures. A total of 15 children (12%) developed premature physeal closure (PPC). The highest incidence of PPC was seen in SH-II injuries, in 10/40 (25%) patients, accounting for 67% in this cohort. Salter Harris-I and Tillaux fractures had no PPC. SH-III and SH-IV had 2 each (13%) and triplane fractures had one patient (7%) with PPC. When the mechanism of injury was considered, the incidence of PPC was highest at 86% (6/7) in motor vehicle accidents (MVA), indicating that these high energy injuries have significantly higher risk of developing PPC as compared with low-energy injuries due to sports or falls. The authors also found that there was a significant linear relationship between the amount of initial displacement and PPC, with a risk of PPC by a factor of 1.15 for every millimetre of initial displacement. It was clearly noted by the authors that the incidence of PPC increased with each attempt though it failed to reach statistical significance.

Comments

In SH-II injuries the area of physis at risk of damage during displacement or reduction is wide compared to other types. Hence the extent of initial displacement, attempts at reduction and the degree of residual displacement, affect the occurrence of PPC. SH-II is often considered as a benign injury and may be treated by a junior member of staff which may increase number of attempts to reduction. With the data from this article SH-II has five times the increased risk of PPC compared to the whole group. If necessary it may be better to open the fracture and do an anatomical reduction instead of multiple attempts at closed reduction.

NEONATAL FOOT DEFORMITIES AND THEIR RELATIONSHIP TO DEVELOPMENTAL DYSPLASIA OF THE HIP - AN 11 YEAR PROSPECTIVE, LONGITUDINAL OBSERVATIONAL STUDY.

R. W. Paton, Q. Choudry
J Bone Joint Surg [Br]. 2009;91-B:655-8.
Investigation performed at the Royal Blackburn Hospital, Blackburn, England

The authors did a prospective observational study in children with various foot deformities. The patients comprised of 614 children with various foot deformities born in Blackburn district, England. Of these, 436 had postural talipes equino varus (TEV), 60 had CTEV, 93 had congenital talipes calcaneo valgus (CTCV) and 25 had metatarsus adductus. These children were assessed clinically and with ultrasonological examination done before 10 weeks of age for diagnosing DDH. Of these 614 children only one hip was of Graf type-III found in a child with postural TEV. Graf type IV hips were observed in eight hips. Six of these were found in association with CTCV and one each with postural TEV & metatarsus adductus.

From this data, statistically CTEV and postural TEV do not have any risk of having DDH while CTCV has a higher risk of having an associated DDH (1:5). Though metatarsus adductus had a small sample size, there was still a case of Graf type VI DDH suggesting an increased severity. The relative risk of having a DDH was 14 times more in a neonate with CTCV than one with CTEV and the relative risk of having a type VI DDH is 17.4 times more in a neonate with metatarsus adductus as compared to the one with postural TEV.

Comments

It is important to accurately know the risk factors if selective ultrasound screening of the hips is practised for neonates. The authors have made an attempt to correlate the deformities of the feet with DDH and observed that CTCV and to lesser extent, metatarsus adductus, carried significant risk of being associated with DDH. Hence neonates with these deformities should be considered for ultrasound screening of the hips.

However the recommendation that postural and fixed CTEV need not be considered as a risk factor, requiring routine screening by ultrasound, has to be taken with caution. This is because 21 type-II and two type-III & IV dysplasias were seen in 496 children, which is approximately one in 25 children. Considering the type II & III hips which are required to be monitored sequentially by Graf's method, it is not justifiable to suggest ignoring this category for screening.

FAMILIAL TUMORAL CALCINOSIS

Harish P and Vrisha Madhuri

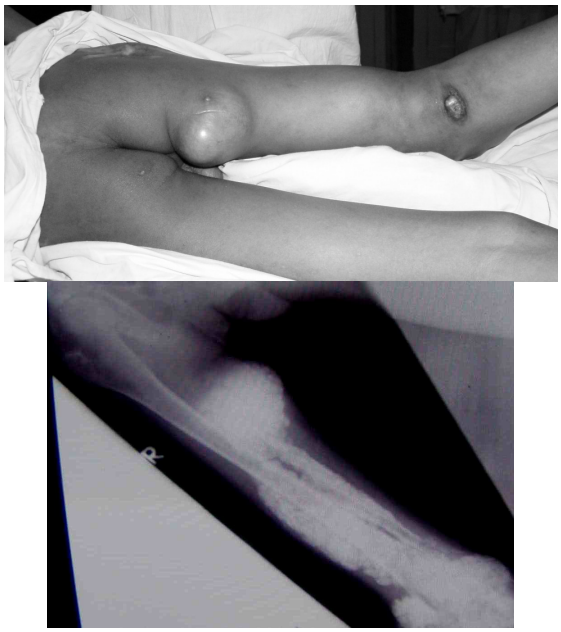
The answer to the previous issue quiz is Familial tumoral calcinosis. Familial Tumoral calcinosis (FTC) refers to a heterogeneous group of inherited disorders caused by a hereditary metabolic dysfunction of phosphate regulation.

On the basis of pathogenesis Smack et al have classified it as: *(a)*Primary normo-phosphatemic tumoral calcinosis, *(b)*Primary hyperphosphatemic tumoral calcinosis and *(c)*Secondary tumoral calcinosis. Hyper-phosphatemic FTC has been shown to result from mutations in three genes: fibroblast growth factor-23 (FGF23), coding for a potent phosphaturic protein, KL encoding Klotho, a co-receptor for FGF23, and GALNT3, which encodes a glycosyltransferase responsible for FGF23 O-glycosylation. Normophosphatemic FTC is associated with absence of functional SAMD9, a putative tumour suppressor and anti-inflammatory protein. [1]

Systemic calcification occurs because the solubility of phosphorus and calcium in the plasma is exceeded. This is believed to happen when the product of plasma calcium and plasma phosphorus is more than 70. [2]

This commonly occurs in the 1st and 2nd decade of life and is characterized by massive calcium deposits usually in the cutaneous and subcutaneous planes in paraarticular locations. Lesions, when large, can ulcerate. These deposits usually occur on the extensor aspects as these sites are subjected to repeated trauma or friction, the commonest being the greater trochanteric bursa. The other sites are elbow, spine, sacrum, hand, and knee. Hyperostosis and diaphysitis are described to be associated with FTC.

Harish P, Fellow in Paediatric Orthopaedics,
Vrisha Madhuri, Prof & Head
Paediatric Orthopaedics Unit
Christian Medical College, Vellore – 632 004
Tamilnadu, India
Email: madhuriwalter@cmcvellore.ac.in



Clinical picture (above) of posterior aspect of thigh and knee showing swellings of calcified deposits in the subcutaneous tissue. Radiograph (below) show extensive calcium deposition in the thigh

Radiographs show a typical appearance of amorphous, cystic, and multilobulated calcification located in a periarticular distribution mainly in the extensor aspects. There can be septae dividing the calcified deposits, described as *chicken wire septae.* Absence of erosion or osseous destruction by adjacent soft-tissue masses is another distinguishing finding of tumoral calcinosis. Axial CT better delineates the calcific mass and it appears as a cystic mass with fluid levels termed the *sedimentation sign*. The lesions with reduced metabolic activity may appear homogeneous and these have a lower likelihood of growth.[3]

The differential diagnoses are dystrophic calcification conditions observed in connective tissue disorders like scleroderma and dermatomyositis (calcinosis universalis or calcinosis circumscripta), in neoplastic diseases like synovial sarcoma, extra-osseous osteosarcoma and

chondrosarcoma, in metaplastic conditions like synovial chondromatosis and in degenerative conditions like calcium pyrophosphate deposition disease, calcific tendonitis and bursitis. Other metabolic conditions which result in soft tissue calcification are also to be differentiated. These include i) Hyperphosphatemia in chronic renal failure ii) Hypercalcemia due to primary hyperparathyroidism, hypervitaminosis D, milk-alkali syndrome, sarcoidosis and hydroxyapatite disease iii) Hyperuricemia causing tophaceous gout.

Surgical excision of the lesions has been well established and is indicated in large and any symptomatic lesions. Local recurrence tends to occur when circumscription is poor. Lesions can occur in different sites after excision and they can ulcerate when large.

Medical treatment is phosphate deprivation using phosphate binding agents orally such as aluminium hydroxide. Yamaguchi et al have reported that Acetazolamide when given in conjunction acts synergistically and reduces the phosphate levels.[4]

Phosphorus binders are most effective when given with food. Binders containing aluminium hydroxide are especially efficient, but calcium carbonate is an effective alternative and may be preferred if there is a need to treat concomitant hypocalcemia. Also systemic steroids and radiation therapy have been shown to be effective.

The patient described in the quiz underwent excision of the lesions around the knee and posterior aspect of thigh. His medical treatment involved long-term aluminium-hydroxide.

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Prof. Vrisha Madhuri, Head, Paediatric Orthopaedics,

Christian Medical College, Vellore 632 004

Tamilnadu, India.