

## POSITIVE

Official newsletter of the Paediatric Orthopaedic Society of India

#### **Editorial**

POSITIVE was first started in 1996 as a scientific newsletter representing the POSI academia. It was a very erudite newsletter which brought current Paediatric Orthopaedic concepts and debates to the fore. As I take over as the new editor for this journal and walk into the footsteps of doyens of Paediatric Orthopaedics like Dr Benjamin Joseph, I hope to enlarge the scope of this newsletter as was originally intended and move to give wider representation to the work done by the Indian Paediatric Orthopaedic community. This requires the participation of the Paediatric Orthopaedic society members and I request all POSI members to contribute original articles and case reports to the newsletter.

The next issue of this newsletter will be brought out in September. All those desirous of contributing to the issue should keep a deadline of 30th June to send the manuscripts to me. The format of the contributions should be as followed by the Journal of Paediatric Orthopaedics. If there is any announcement which needs to be circulated such as scientific meetings, calls for participants in research studies, fellowship announcements, kindly hand them in by the end of August.

I am grateful to Dr Kaye E Wilkins and Dr William G Mackenzie for their contributions to this issue. The present issue is largely the effort of staff and fellows of Paediatric Orthopaedic department at Christian Medical College Vellore. I earnestly request members of Paediatric Orthopaedic community to join the list of contributors.

Vrisha Madhuri.

Vaishe Hadhimi

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March '09

#### PRINCIPLES OF FRACTURE REMODELING IN CHILDREN

Dr. Kaye E Wilkins

There are two advantages in treating children's fractures. First, the healing process is very rapid. Non union is a rare event in the pediatric age group (20). The second advantage is that there is a very good remodeling capacity should there be less than anatomic alignment of the affected bone once the fracture has healed. Any individual treating fractures in the pediatric age group should have a full understanding of how pediatric fractures heal and how the remodeling process occurs.

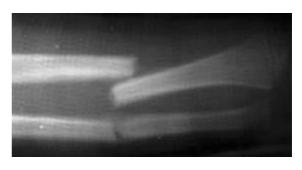
#### THE BASIC BIOLOGY OF FRACTURE HEALING

Healing of fractures in children has been described by two authors (15, 16) as occurring in three phases: 1) Inflammatory, 2) Reparative and 3) Remodeling. Much of the following discussion of each of these processes has been taken from their work (15, 16).

#### Inflammatory

The initial phase is one of acute inflammation following the loss of integrity of the osseous structure (Figure 1A). This is the result of the hematoma formation that occurs from the rupture of the blood vessels. This hematoma contains a lot of fibrin which is rapidly replaced by a collagen scaffold. This collagen scaffold serves as the lattice framework for the initial woven bone. The hematoma also incites the production of proteins which stimulate the differentiation of fibroblasts. cells into chondroblasts. osteoblasts and angioblasts all of which are essential for the formation of new bone.

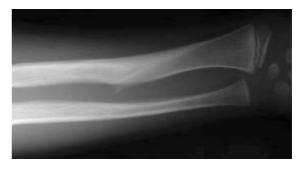
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Figure 1. Phases of fracture healing. A. The immediate angulation following the fracture. This represents the hematoma and *inflammatory* phase. B. At eight weeks there is good provisional callus laid down in a random manner. This is the *reparative* phase. C. By eighteen months the bowing has remodeled along stress lines and the angular deformity has been pretty well resolved. This is the *remodeling* phase

#### **Reparative Phase**

It is during this phase that the initial callus is formed. There is first the invasion of the hematoma by the fibrovascular tissue from both endosteal and periosteal sources. Subsequently, new bone forms from both

endochondral (usually from an endosteal source) ossification and intramembranous (usually from the cells of the periosteum) ossification. This early type of bone is laid down haphazardly and in a random manner to fill the gap between the bone ends of the fracture. Because it is weaker structurally, a larger amount needs to be produced, thus the term "Quantity Bone" (Figure 1B). produces the so-called provisional callus which provides temporary stabilization. This callus still does not provide the rigid stability necessary for full physical activity. reparative phase occurs during the first two to three months of the fracture healing process. **Remodeling Phase** 

It is during this phase, which can last months and even years in some osseous structures, that the provisional callus is gradually removed and new bone is laid down along the lines of stress (Figure 1C). Thus we now have the production of "Quality Bone" which is rigid and can support the normal physical activity of the child.

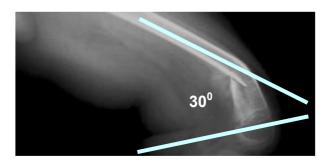
## BASIC PRINCIPLES OF THE REMODELING PROCESS

#### The Location Affects Remodeling

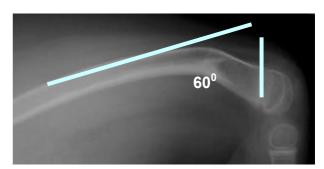
Metaphysis. There is a difference in the rate of remodeling as to the location of the fracture site. The metaphysis serves as an active remodeling area in the development of normal bone growth. It is the area where the quantity of woven bone produced in the adjacent physis is replaced with the more structurally sound quality compact bone of the diaphysis. Thus, this area has already in place an increased vascularity with much more osteogenic potential to facilitate fracture healing. The osteogenesis in this area is normally active.

**Diaphysis.** The diaphysis is primarily an area in which there is relatively dormant osteogenesis. Most of the bone production here is a balance of subperiosteal intramembranous ossification on the surface

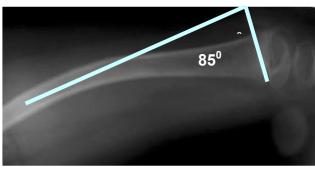
coupled with endosteal bone reabsorption in the medullary canal. The bone here is rigid, compact cortical bone and thus is relatively avascular. As a result, there is less remodeling potential in this area. Fractures in this area take longer to heal and remodel.



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Figure 2. Physeal realignment. A. The appearance two weeks post-injury of a child with severe cerebral palsy. The  $30^{\circ}$  of alignment of the long axis of the shaft with the physis was accepted. B. By nine months the physis has realigned itself to be within  $60^{\circ}$  of the long axis of the shaft. C. At eighteen months there is almost complete realignment of the distal physis with the shaft, the angle now being  $85^{\circ}$ .

### How Does the Remodeling Process Occur?

#### Angulation.

PHYSIS. In the skeletally immature individual 75% of the angular remodeling

takes place in the physis (29). Pauwels (21), Ryoppy Karaharju and (23)have demonstrated that both physes adjacent to a tend to realign to become perpendicular to the forces acting through them by a process of asymmetrical growth. The concave side is stimulated to grow more rapidly to align the physis so as to become perpendicular to the long axis of the shaft of the bone (Figure 2). Once the physis is realigned, it then resumes symmetrical growth.

<u>DIAPHYSIS</u>. In the diaphysis remodeling follows Wolff's law (28). Here there is increased pressure (compression) on the concave side which stimulates new bone formation. On the convex side, the bone is under tension and thus there is reabsorption of the convexity (Figure 3). About 20% of the remodeling of the angulation occurs in this area.

#### Length.

FEMORAL OVERGROWTH. It has been recognized for a long time that the fracture healing process stimulates bone growth (25). This growth stimulation is most prominent with most fractures involving the femoral shaft (Figure 4). Various length amounts have been reported for femoral overgrowth following fractures of the shaft. Probably the most detailed and accurate study was that by Shapiro (24). In his review of seventy-four patients under the age of thirteen, he found the average overgrowth was 0.92 cm. (Range 0.4-2.7). He found the overgrowth to be independent of age, fracture level or the position of the fracture at the time of healing. In the majority of patients there was some effect of growth stimulation for as long as three years and six months post-injury. It is felt that this growth stimulation was due to an increase of blood flow to the adjacent growth areas in response the fracture healing process.

TIBIAL OVERGROWTH. On the other hand, growth stimulation following fractures of the tibial shaft is age-dependent (13). The maximum stimulation of 4.2 mm occurred in

the three to five year age group. In older children, the stimulation was less with some actual growth inhibition as the child reached maturity. There does appear to be a greater tendency to increase tibial overgrowth in those patients with open fractures (5).

#### Rotation.

Remodeling of rotation for practical purposes does not occur (7).

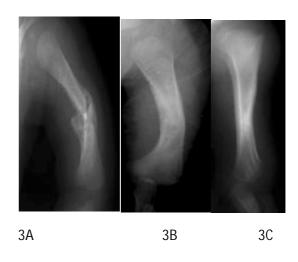


Figure 3. Diaphyseal appositional remodeling. A. Four week old fracture in a neonate showing new bone formation on the concave side and along with early reabsorption on the convex side. B. By eight months there has been considerable remodeling of the shaft. C. At fourteen months there is complete remodeling and realignment of the diaphysis of this humerus.

#### Relationship to Growth Potential.

<u>UPPER EXTREMITY</u>. In the upper extremity the proximal and distal growth centers account for the majority of growth in the extremity. Thus, the remodeling potential in these areas is extensive. Fractures about the elbow, especially the distal humerus, have very little remodeling potential.

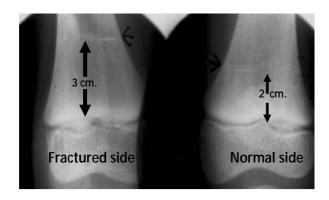
LOWER EXTREMITY. The opposite is true of the lower extremity. If there is any expected remodeling, it occurs to a greater degree in the distal femur and proximal tibia (knee region). However, the amount of

remodeling potential is less than that seen in the upper extremity.

### Overall Factors Affecting the Remodeling Potential

The three major factors which have a bearing on the potential for angular remodeling are:

- 1. Skeletal age.
- 2. Distance to the joint.
- 3. Orientation to the joint axis.



**Figure 4. Femoral overgrowth.** This eight year old male sustained a fracture femur eighteen months previously. X-rays of both distal femurs show that there has been a 3 cm. migration of the Harris-Park growth arrest lines on the fractured side versus only 2 cm of migration these lines on the normal side.

#### REMODELING OF SPECIFIC FRACTURES

The remodeling capacity of fractures in the various parts of the upper and lower extremities varies considerably. One of the best reviews of this subject can be found in the recent article by Gasco and De Pablos (11). In the following section, we will review the remodeling capacity of the various areas and the recommended degrees of angular deformity or shortening which can be expected to remodel into an acceptable cosmetic and functional result.

#### Fractures of the Hand.

**Phalanges.** Very little remodeling can be expected in the fractures involving the distal portions (supracondylar and condylar

areas) of the proximal and middle phalanges(12). In the proximal portions of these proximal and middle phalanges, some remodeling can be expected in the saggital plane. Less can be expected in the coronal plane. Unfortunately, very little information is available in the literature as to the acceptable degrees of angulation for phalangeal fractures in children.

Metacarpals. Most οf the controversy centers in regards to acceptable angulation of apex dorsal fractures of the fourth and fifth metacarpal necks. Some are very liberal in their manipulative indications accepting up to only 35-40° of apex dorsal angulations (2). Others are very conservative and have found that the ultimate function with those fractures which are not manipulated is equal to those who were manipulated (4, 18). The only difference in those that were not manipulated was the absence of the prominence of the metacarpal head (knuckle) in those not manipulated.

A summary of the expected degrees of remodeling of the various fractures of the bones of the hand is presented in Table I.

# Proximal and Middle Phalanges (12) <10 years 20-25° in any plane >10 years 10-15° in any plane Metacarpal Neck

35-40° apex dorsal (2) Others state almost none of these Fractures need to be realigned (4, 18).

## Metacarpal Shaft No good data found

Table I. Acceptable Angular Malalignment of Hand Fractures

#### Fractures of the Distal Radial Physis.

This area has probably one of the greatest potentials for remodeling of any fracture in the immature skeleton (Figure 5). Aitken (1) demonstrated many years ago that these fractures have a tremendous capacity for remodeling. He pointed out that up to

50% displacement of the fracture fragments can be expected to remodel fully if there is at least one and one half years of growth remaining.

#### Fractures of the Distal Radial Metaphysis.

Angular Remodeling. The distal radial metaphyseal fractures likewise have a great potential for angular remodeling. The commonly accepted angulation that will fully remodel with five years of growth remaining is 30-35° in the sagittal plane and 10° in the coronal plane. In many cases however, the remodeling angulation may not be complete but there are no functional or cosmetic residual.







Figure 5. Remodeling of the distal radial physis. A. The appearance of the fracture fragments on cast removal four weeks post-injury. There is marked displacement of early callus formation. B. At three months the fracture has shown considerable remodeling of both the metaphysis and early realignment of the epiphysis. C. By fifteen months there is complete realignment of the distal epiphysis and remodeling of the metaphysis. The nonunion of the ulnar styloid was asymptomatic.

**Bayonet Remodeling**. Bayonet apposition can be expected to remodel in patients up to twelve years as long as the linear alignment is nearly anatomic (30) (Figure 6).

**Reduction Necessary?** In a recent report Do and associates (8) felt that the degree of remodeling in this area was so great that the majority of their distal radial metaphyseal fractures did not even require a primary reduction. They accepted up to 15° of primary angulation and 1 cm. of shortening in boys up to fourteen years of age and girls up to twelve years of age. It was their opinion that it was a waste of time and financial resources to do a manipulation on these displaced fractures meeting these criteria. It was their observation that even those fractures that were completely unreduced had essentially complete remodeling at the termination of their growth.



Figure 6-a

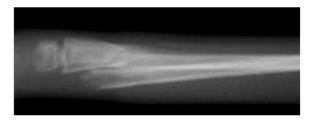


Figure 6-b

Figure 6. Bayonet remodeling. A. Position of the fracture fragments show early callus formation. This was the best position of the fracture fragments that could be achieved by closed reduction. B. At eighteen months there has been essentially complete realignment of the distal metaphysis with the proximal metaphysis. This patient had a full range of forearm and wrist motion.

#### Fractures of the Radial and Ulnar Shafts.

Multiple Factors. Acceptable malalignment of the shafts of the radius (and ulna as well) is dependent on multiple factors. Based upon his wide experience with these fractures, Price (22) has set down some guidelines for the various factors involved (Table II). He also found that impingement across the interosseous space by the fracture

fragment was an unpredictable factor in determining the ultimate outcome.

Age	Angul- ation	Malro- tation	Displace- ment	Loss of radial bow
>	15°	45°	Complete	Yes
9yrs				
<	10°	30°	Complete	Partial
9yrs				

Table II. Acceptable Malalignment for Radial Shaft Fractures.(22)

**Plastic Deformation.** In a study by Vorlat and De Boeck (26), it was found that over the age of six years greater than 10° of plastic deformation of either the radial or ulnar shafts would result in an unacceptable result.

#### Fractures of the Radial Neck.

**Angulation.** There is not a total consensus on the amount of angulation of a radial neck fracture that can be accepted with resultant satisfactory remodeling. The most commonly accepted number is  $30^{\circ}$  of angulation (2, 6). However, other studies have demonstrated that even those with up to  $50^{\circ}$  of angulation can be expected to achieve good results.

*Translocation.* Originally it was felt that as little as 2 mm of translocation results in a poor outcome. However, more recent studies have shown that up to 5mm of translocation will remodel (6).

Clinical Examination. Probably the best method of determining what degree of deformity will result in an acceptable outcome involves a clinical examination under sedation or anesthesia to determine the passive range of forearm motion. If there is at least 50° of supination and 50° of pronation, the patient should be expected to have a satisfactory functional result.

## Supracondylar Fractures of the Distal Humerus.

**Angulation** – **No.** Very little angulation in the *sagittal* plane can be expected to remodel. Up to loss of 20° of the

shaft-condylar angle can be tolerated. This is usually only manifest as a lack of full elbow flexion with some increase in elbow hyperextension. In the *coronal* plane, no angular remodeling can be expected. Angulation into varus will result in an unacceptable cosmetic deformity. It has also been shown that cubitus varus produces some functional effects such as recurrent fractures of the lateral humeral condyle or late ulnar nerve neuropathy (30).

Translocation – Yes. On the other hand, translocation of as much as one hundred percent in either plane has been shown to demonstrate complete remodeling (30).

#### Fractures of the Humeral Shaft.

There is considerable remodeling of the humeral shaft especially in the very young age groups. A good example of the remodeling capacity in the infant is seen in Figure 3. Kwon and Sarwark (19) have reviewed the literature and come up with some guidelines as to acceptable displacement (Table III). Fortunately, minimal angulation is well hidden by the muscles of the arm.

Table III. Acceptable Displacement for Humerus Shaft Fractures (19)

Type of Displacement	Amt.
Varus Angulation	20-30°
Apex Anterior Bowing	20°
Loss of Internal Rotation	15°
Bayonet Shortening	2 cm.

#### Fractures of the Proximal Humerus.

Proximal Physeal Injuries. Because of the presence of fractures through the proximal humeral physis tend to develop angular deformities because of the presence of the rotator cuff muscles acting only on the proximal fragment. Opposing muscles are still attached to the distal fragment. Fortunately however, because of the marked flexibility and circumduction nature of the shoulder both considerable displacement and angulation can be tolerated in the younger age group. Beaty (3) has set out guidelines for each age group (Table IV). In those patients that use their upper extremities for high performance athletic activities less than a near anatomic reduction however may result in some loss of athletic performance.

Age	Displacement (degrees,%,)		
< 5 yrs	Up to 70° angulation, 100% displacement		
5 – 12 yrs yrs	40-70° angulation		
>12 yrs	Up to 40% displacement, 50% apposition		

Table IV. Acceptable Displacement for Proximal Humeral Fractures (3)

Proximal Metaphyseal Injuries. With this fracture pattern there is some resistance to external rotation and abduction of the proximal fragment because of the persistence of muscle insertion of the pectoralis major on the proximal fragment. Thus many of these fractures if complete, tend to present with bayonet apposition. By and large this bayonet apposition can be expected to remodel to a satisfactory degree if there is at least two years of growth remaining.

#### Fractures of the Femoral Shaft.

Angular Malalignment. Malalignment in both sagittal and coronal planes is somewhat age dependent. Kasser (17) has outlined very nicely his recommendations for acceptable angulation in the various planes as it relates to the specific age groups (Table V). Because of the normal natural anterior bow of the femur, more angulation can be tolerated in the sagittal plane.

**Shortening.** The amount of shortening expected to correct has been discussed

previously in this section on femoral overgrowth. It must be remembered that a combination of angulation with shortening has an additive effect and can spell trouble regarding an acceptable outcome (Figure 7).

Loss of Rotation. As mentioned previously, Davids (7) has shown that rotation does not significantly remodel. He did find in his studies that up to 25° can be well tolerated.

Deformity	<8yrs	>8yrs
Valgus	5°	5°
Varus	10°	5°
Anterior Angulation	10°	5°
Posterior	5°	00
Shortening	10 mm.	5 mm.
Rotation	5°	5°

Table V. Acceptable Angular Malalignment For Fractures of the Femoral Shaft (17)

#### Fractures of the Tibial Shaft.

Angular Malalignment. The tibia is very unforgiving in its ability to remodel. This may be because it is composed of a very large amount of diaphyseal bone. Remodeling in the sagittal plane is better than in the coronal plane. Varus has a better chance to remodel than valgus (14). Heinrich has set out some good guidelines for the remodeling potential of tibial shaft fractures according to the patient's age (Table VI) (14).

*Translation.* Because of the subcutaneous nature of the tibial shaft there may be concern of the effect of translocation on the clinical appearance. One hundred percent translocation will result in a satisfactory outcome in the young child, whereas in the adolescent the goal should be to achieve at least 50% apposition (14).



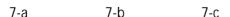


Figure 7. Combined deformities equals trouble. A. A six year old female who is five weeks postinjury shows 4 cm. of shortening on the A/P radiograph. B. On the lateral radiograph, in addition to the shortening there is 33 degrees of apex anterior angulation. C. At three years the combined angulation and shortening have produced a resultant 3.5 cm. discrepancy which has persisted and not changed over the last year. This shortening resulted in both unacceptable functional and cosmetic results.

Table VI. Acceptable Angulation of Malalignment for Fractures of the Femoral Shaft (14)

Age	Varus/Valgus	Sagittal
Birth-2yrs	30°	30°
2-5yrs.	15°	20°
6-10 yrs.	100	15 <b>º</b>
11yrs +	50	100

Fortunately for the treating surgeon, children have a tremendous capacity to remodel malalignment of their fractures should it occur. *This is no excuse for the treating surgeon not to make every attempt to obtain as anatomical an alignment as possible.* If this cannot be achieved by conservative methods, then serious consideration should be given to achieving a satisfactory reduction by operative means.

#### **SUMMARY**

In treating fractures in children, the surgeon must have a good knowledge of the three phases of bone healing, i.e. inflammatory, reparative and remodeling and

understand how they contribute to the final recovery of the fracture healing process. By and large the ability to remodel is dependent upon the bone involved, the patient's age, the proximity to the joint and its orientation to the joint axis. In the typical long bone, 75% of the remodeling occurs by reorientation of the physis while appositional remodeling of the diaphysis can be expected to contribute only 25% to the remodeling process.

The various values of acceptable alignment for each of the major fracture patterns listed should serve only as guidelines. The patient's functional capacity and the surgeon's experience should also be factors in determining whether to depend upon the remodeling capacity of the specific fracture or to consider performing a more aggressive, invasive technique to achieve a satisfactory result.

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#### Announcement

## 16<sup>th</sup> annual conference of POSI (POSICON 2010)

Theme: "Reconstruction Procedures in

Paediatric Orthopaedics (Foot & Hip)" Dates: 23<sup>rd</sup> and 24<sup>th</sup> January 2010

Venue: Gandhi Medical College, Hyderabad

#### 3<sup>rd</sup> POSNA-POSI workshop

Dates: 22<sup>nd</sup> January 2010 (Pre-conference Workshop)

24<sup>th</sup> -25<sup>th</sup> January 2010 (post-conference Workshop)

Venue: Gandhi Medical College, Hyderabad

- a. Basic Hip Course (Paediatric Hip Disorders) (22<sup>nd</sup> January,2010)
- b. Reconstruction of long bone defects in children(22<sup>nd</sup> January,2010
- c. Conservative management of Club foot (22<sup>nd</sup> January,2010)
- d. Advanced Hip Course (Adolescent Hip Disorders) (2<sup>4th</sup> -2<sup>5th</sup> January,2010)

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#### THE SPINE IN CHILDREN WITH SHORT STATURE

#### William G Mackenzie

The spine is almost always affected in disorders causing short stature in children. The various spine problems in skeletal dysplasia are as follows:

- 1) Instability This is usually Atlanto-axial.
- 2) Stenosis This could be at the cervical, thoracic or lumbar level.
- 3) Deformity This could be scoliosis, kyphosis or lordosis or a combination of these deformities.

#### **ACHONDROPLASIA**

This is one of the most common dysplasias with a variety of spinal problems presenting at different ages.

- A) <u>Cranio-cervical stenosis</u> (foramen magnum stenosis) due to hypoplasia of foramen magnum is common in achondroplasia but not in other types of skeletal dysplasia. It causes Cervicomedullary compression which can lead to
  - developmental delay
  - hypotonia
  - central sleep apnea due to cord compression and should be differentiated from obstructive type of sleep apnea usually a result of mid face hypoplasia.
  - feeding difficulties
  - hemiplegia, quadriplegia
  - sudden death usually in children < 1 year old.

#### Evaluation-

- MRI (AAP recommendation for MRI in perinatal period)
- Sleep study

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

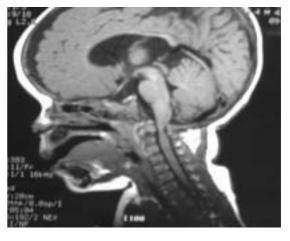


Figure-1. Cervicomedullary compression in achondroplasia

Natural history – There can be a spontaneous relief of compression over first 3 years of life. Apnea monitoring is important to avoid disasters.

Surgical decompression- It can be a difficult decision to decide for or against surgery. Cranio-cervical decompression is all that is needed and fusion is not required.

Significant complications include altered CSF dynamics and neurological injury. Shunts are commonly needed. Good results are seen in symptomatic children but the role of prophylactic surgery is still debatable.

B) <u>Thoracolumbar kyphosis</u> is a developmental problem which is mild at birth and progresses due to unsupported sitting by hypotonic infants with achondroplasia. More than 90% resolve when child starts walking.

#### Treatment

a) Bracing – Stiff kyphosis >50° at more than two years of age with progressive anterior vertebral wedging requires bracing. Usually a thoraco-lumbar-sacral orthosis is prescribed. Some authors suggest early bracing.

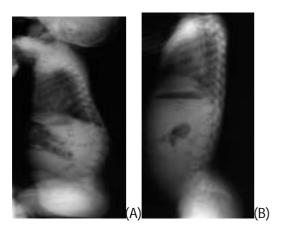


Figure 2 A & B -spontaneous correction of thoracolumbar kyphosis

b) Anterior and posterior spinal fusion with instrumentation – Indications-Residual stiff kyphosis >40° at or beyond five years of age with anterior vertebral wedging and spinal stenosis symptoms. Incidence of neurological damage is higher because of diminished spinal canal volume.

C) <u>Lumbar spinal stenosis</u>: The spinal canal volume is 1/3 of normal. The stenosis occurs as a result of short pedicles, interpedicular narrowing, thick laminae and disc protrusion and is aggravated by lordosis (sway back).

Symptoms include low back pain, numbness, weakness, incontinence, neurogenic claudication relieved by squatting or sitting. With progression, walking endurance is reduced and neurological signs like, clonus, hyperreflexia and lower extremity weakness may develop. One third of the children are symptomatic by 15 years, and 91 % are symptomatic by 30 years of age. Evaluation is by MRI, CT myelogram. Treatment-

1. Non-surgical- Weight loss, lumbar flexion exercises, bracing to reduce lordosis

#### 2. Surgical-

- Hip extension osteotomy during lengthening.
- Wide posterior decompression- usually T-12 to S1. The decompression should

extend at least 3 levels cephalad to the myelographic block and distally till S2 and laterally at least to facets but preserving the facet joints. As instability rarely develops primary fusion is not necessary unless kyphosis is present. Due to the nature of the disease dural tears are a common complication during decompression.

Neurological deficits of short duration are usually reversible with surgical decompression. A recurrence of deficits requires additional decompression. Remember that cervical and thoracic stenosis can occur in achondroplasia. Upper cervical stenosis without instability is seen in Metatropic dysplasia and requires posterior decompression and limited fusion.

#### **CERVICAL INSTABILITY**

Atlantoaxial Instability is common in a number of dysplasias. It is not a feature of Achondroplasia. Some of these dysplasias are:

- Spondylo-epiphyseal Dysplasia and variants
- Morquio's Syndrome
- Pseudoachondroplasia
- Metatropic Dysplasia
- Kniest Dysplasia
- Metaphyseal Chondrodysplasia

The instability is usually due to:

- Ligamentous laxity
- Odontoid hypoplasia/aplasia
- Os odontoideum

Associated stenosis is quite common.

The presenting features are those of cervical myelopathy and these include persistent hypotonia, apnea, delayed motor development, decreased endurance and quadriplegia. On examination patient has weakness, hyperflexia, clonus, abnormal proprioception and vibration sense. Children with SED present earlier than those with Morquio's syndrome.

The radiological evaluation consists of:-

- a) Cervical spine lateral radiographs in Flexion and extension.
- b) Flexion/extension MRI
- c) CT scan -to assess C1 arch, anatomy for C1-2 screw.

The other investigations carried out include urodynamics. A re-evaluation is needed yearly as developmental instability can occur.

#### Treatment

Cervical Fusion is indicated for:

- a) Instability without cord compression -ADI > 8 mm or SAC < 13 mm
- b) Instability with cord compression If the instability can be reduced relieving cord compression then fusion can be done in the reduced position.

Decompression combined with cervical fusion is required when there is cervical myelopathy or for instability with cord compression which cannot be reduced.

Fusion is extended to occiput if:

- a) Occipito-atlantal instability exists.
- b) C1 midline synchondrosis is large after C1 decompression.
- c) Inadequate posterior elements or the surgeon is unable to achieve C 1-2 stability.

Surgery requires fibre optic intubation and neurophysiologic monitoring. C 1-2 Fixation is done by transarticular screws or by Brookes wiring.

Immobilization after Surgery – Minerva cast, Halo vest or cast, or cervicothoracic orthosis are usually worn till 12 weeks.

Pseudarthrosis is uncommon in fusions with instrumentation.

#### **CERVICAL KYPHOSIS**

This is seen in

- Diastrophic Dysplasia
- Camptomelic Dysplasia
- Larsen's syndrome



Figure 3 Cervical kyphosis in larsen's syndrome

Treatment: - spontaneous improvement often occurs by age 5 in diastrophic dysplasia

Brace- treatment is indicated in children with progressive kyphosis with no instability, deformity of less than 60 degrees in a child aged four years or more and with normal neurological function.

Surgical treatment is indicated in a child with signs and symptoms of cord compression or, severe or progressive kyphosis despite bracing. surgery The choice of is anterior decompression, fusion (strut grafting) and posterior spinal fusion. One needs to be aware during surgery of cervical spina bifida which is common. After posterior fusion progressive correction of kyphosis occurs. decompression is required prior to fusion in case of cord compression.

#### **Scoliosis**

This is commonly seen in spondyloepipyseal dysplasia congenita, pseudoachondroplasia and diastrophic dysplasia.

Treatment strategies are the same as for idiopathic scoliosis consisting of brace therapy for mild curves and surgical anterior and posterior fusion for severe curves. Usually

curves more than 50 degrees require surgery. Growing rod systems may be indicated in young children.

**KYPHOSCOLIOSIS** - is seen commonly in

- Chondrodysplasia Punctata,
- Metatropic Dysplasia

Severe curves are seen and surgical management is quite difficult. Early

management is needed. It is a progressive deformity and associated with severe restrictive lung disease.

This article is based on the lecture delivered at the skeletal dysplasia symposium, annual meeting of Paediatric Orthopaedic Society of India, Jaipur, 2007.

#### **Announcement**

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Theme Paediatric Orthopaedics Interested surgeons may contact.

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#### <u>POSI-Dr.Kameshitha Young Surgeon's Forum</u> <u>Prize</u>

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You may down load the application form the POSI web site <a href="www.posi.in">www.posi.in</a> or contact the Secretary for one.

Last Date for sending the application: 31<sup>st</sup> June 2009.

#### MULTIFOCAL SYNCHRONOUS OSTEOSARCOMA A CASE REPORT

#### Harish P and Vrisha Madhuri

#### **ABSTRACT**

Multifocal Osteosarcoma is a rare entity. Typically there is a dominant symptomatic primary lesion in one of the bones, with multiple symmetrical and asymptomatic lesions in other bones and no pulmonary metastases. The prognosis for such a sarcoma is very poor despite aggressive chemotherapy and resection of tumor.

Osteopoikilosis is a rare osteosclerotic dysplasia, characterized by multiple osteosclerotic lesions in metaphysis and epiphysis. Majority are asymptomatic and are usually found incidentally. Osteopoikilosis should be differentiated from multifocal osteosarcoma or osteoblastic metastasis especially when the presentation is atypical and also when the lesions are symptomatic.

We present a case report of an eight year old girl who presented to us with pain in the region of right knee of short duration. Initially a diagnosis of osteopoikilosis based on the radiological findings was considered. However, biopsy of the symptomatic lesion of right femoral condyle showed high grade osteosarcoma. Technetium-99 MDP bone scan (Tc-99m) showed increased uptake at multiple sites which correspond to the lesions in the radiographs. Hence, osteopoikiolosis with atypical presentation should be differentiated from osteoblastic secondaries and synchronous multifocal osteosarcoma. Bone scan and biopsy would aid in establishing the diagnosis.

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#### INTRODUCTION

Multiple sclerotic osteosarcomas, socalled "multifocal osteosarcoma" "osteosarcomatosis," is an uncommon entity first described in 1883. <sup>1</sup> The reported incidence ranges from 1% to 10% of osteosarcoma.<sup>2</sup> By definition, it is the occurrence of the tumour at two or more sites in a patient with or without pulmonary metastases and may be synchronous or metachronous.<sup>2</sup> Whether these tumors are multicentric in origin or are bone metastases from a symptomatic primary osteosarcoma was a matter of debate. But now, many consider it as metastatic.<sup>3,4</sup> On radiographs, they are seen symmetrical smudgy lesions in metaphysis of long bones, vertebrae and pelvis. In the literature, these multiple skeletal lesions have been reported to occur in young patients and are described as symmetric and arising at the time of or within a few weeks after a radiographically symptomatic, dominant osteosarcoma is diagnosed.<sup>2</sup> Simultaneous detection of such skeletal lesions with the primary tumor is designated as synchronous multifocal sclerosing osteosarcoma. In contrast, lesions may appear later, at different intervals or after treatment of the primary tumor. These lesions are designated as *metachronous* tumors<sup>2, 5</sup>.

Osteopoikilosis is a heritable condition, usually asymptomatic, in which osteosclerotic dysplasia of bones develops during childhood and persists throughout life.<sup>6</sup> The reported prevalence is less than 0.1 per million<sup>6</sup> to 1/50000<sup>7</sup>, and it is inherited as an autosomal dominant condition.<sup>6</sup> The diagnosis is usually made incidentally on radiographs which show symmetrical round or oval areas of spotted condensation in the spongiosa involving multiple bones predominantly in the bones of hand and feet and in ends of long bones and

pelvis.<sup>6</sup> Most often it occurs without symptoms and several members of the family may be involved when screened for.<sup>6</sup> It occurs in both sexes and has been reported in every bone.<sup>6</sup> Under microscope they are seen as focal condensations of compact lamellar bone interspersed within cancellous bone.<sup>6,8</sup> The lesions as such do not require any intervention and have good prognosis.

Complications occurring osteopoikilosis are very rare. Only one report of osteosarcoma associated with osteopoikilosis has been reported.9 Very rarely cases of malignant change in a case of osteopoikilosis have been reported. 9,11,12 The main challenge lies in differential diagnosis from other similar conditions like osteopathia striata. melorheostosis, and more importantly multifocal osteosarcoma in pediatric age group and osteoblastic secondaries in adults. Unlike osteosarcoma, osteopoikilotic lesions do not show any uptake in Tc-99m bone scan. 10, 13,14

Some patients may have features of melorheostosis or osteopathia striata in association with osteopoikilosis. The coexistence of two or more sclerosing conditions mixed as sclerosing dysplasia.<sup>7,14,15</sup> The issue of differentiation of osteopoikilosis from multicentric osteosarcoma arises when there is a symptomatic lesion or the presentation is atypical. This is important for management and prognosticating the condition. We report an eight year old girl who presented with pain in the region of the right knee. Initial radiographs were suggestive of osteopoikilosis but biopsy of the symptomatic lesion showed high grade osteosarcoma.

#### **CASE REPORT**

An eight year old girl presented with complaints of vague pain in the inner aspect of right knee joint region for two to three months, which was preceded by a trivial injury from a fall. She had right sided mild limp on the right side associated with pain after walking a short distance. The child was well built and nourished and had no loss of weight or appetite. Systemic

examination was normal. She had an antalgic gait on the right side. There was tenderness and thickening over right medial femoral condyle. There was no effusion or fixed deformities of the right knee joint and she had movements ranging from full extension to 130 degree of flexion. The rest of the musculoskeletal examination was normal.

Plain radiographs showed diffuse sclerosis of the distal half of the femur extending from the physeal plate to the proximal diaphysis similar to skip lesions seen as areas of patchy sclerosis. The medial aspect of distal metaphysis of right femur showed areas of ill-defined destruction of bone and periosteal reaction. Punctuate round to oval osteo-sclerotic lesions were also seen in the distal epiphysis of the femur on the medial side. Similar round to oval osteo-sclerotic lesions as well as smudgy sclerotic lesions were noted in the metaphysis of long bones of the lower limbs, distal ulna, proximal humerii and vertebral bodies of the lumbar spine.

Laboratory investigations revealed an elevated ESR of 42 mm at the end of 1st hour. Since the right femur lesion was rapidly progressive showing some areas of irregular destruction and periosteal reaction, a malignant process was suspected. Needle biopsy taken from right distal femur was reported as high grade osteosarcoma. Screening for metastases was done with Tc-99m bone scan and high resolution CT of thorax and abdomen. Technetium-99 MDP bone scan showed an abnormal diffuse linear increased tracer activity in the entire length of right femur and proximal half of both humerii. Epiphyseal ends of long bones of both lower limbs and both sacroiliac regions showed increased tracer uptake. Foci of increased tracer uptake were also seen in the skull, ribs and multiple vertebrae. Biochemical tests included serum alkaline phosphatase(ALP) and lactate dehydrogenase(LDH) which were in excess of 6800 and 2500 units respectively. Serum calcium and phosphorus levels were normal.

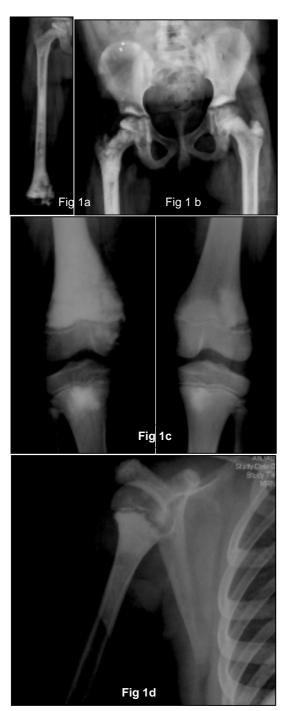


Fig 1a-d: Plain Radiographs showing osteosclerotic lesions involving the distal half of right femur with cortical destruction in the medial femoral condyle. Smudgy osteosclerotic lesions are seen in metaphyses of femur, tibia, proximal humerus

With the report of these investigation as background, a diagnosis of multifocal synchronous osteosarcoma was considered. The child was planned for immediate treatment with aggressive chemotherapy. At present child is undergoing treatment at a cancer institute elsewhere of her parents choice.

#### **DISCUSSION**

Review of literature for osteosarcoma with metastases and osteopoikilosis with osteosarcoma led us to consider a diagnosis of multifocal synchronous osteosarcoma. Multifocal osteosarcoma constitutes about 1-10% of osteosarcoma patients.<sup>2</sup> They often present as a single symptomatic lesion with asymptomatic lesions at other sites commonly in the metaphysis of long bones with or without pulmonary metastasis. Radiologically they are seen as smudgy osteosclerotic lesions in the metaphysis of long bones, pelvis and sometimes in vertebrae. 16 Our child was apparently normal till about 3-4 months prior to presentation. She had symptoms only in the region of right knee but did not have any symptoms in the rest of the areas where the radiographs showed lesions. As the lesions were similar to osteopoikilosis especially in the right femur, we were misled initially to consider the generalized skeletal lesions as osteopoikilosis. However the literature shows that osteopoikilosis when present as an isolated entity are often asymptomatic and are usually discovered when radiographic screening is done for some other reason.<sup>6,7</sup> But the symptomatic ones are often mixed variety of osteosclerotic dysplasia and those with complications.<sup>7,9,13-15</sup> Only three reports of tumor associated with osteopoikilosis have been described in the literature. 9,14,15 Only one of these was an osteosarcoma of the proximal tibia in a 42 year old lady. 9 The other two lesions were chondrosarcoma and giant cell tumor. 14,15 In the previous reported case of osteosarcoma the underlying osteopoikilosis was confirmed by histological examination of other nonmalignant lesions form the resected specimen.9

Since the child had pain and radiographic evidence of bony destruction, all of which are atypical of osteopoikilosis, malignancy with metastases was suspected. In such a situation tissue diagnosis with biopsy was thought to be crucial in arriving at the definitive diagnosis. It showed features of high grade osteosarcoma.

The single dominant symptomatic lesion of right distal femur was proven to be osteosarcoma by biopsy in this patient. The major consideration then was regarding the other lesions. Were they metastasis or osteopoikilosis? Asymptomatic simultaneous occurring lesions at other sites were found at presentation. Grossly elevated serum Alkaline Phosphatase and LDH levels, along with bone scan picture of diffuse increased uptake in right femur, ends of left femur and upper ends of tibiae and humerii, supported a diagnosis of multifocal osteosarcoma.

The possibility of osteopoikilosis associated osteosarcoma as a differential was rejected because many of the lesions were smudgy and almost all except the very small ones had an increased uptake on bone scan. These are not osteopoikilosis features. The lack of any lesions in the hands and carpus was against a diagnosis of osteopoikilosis. Some of the lesions in both femora and distal ulna were similar to that are seen in osteopoikilosis as they were discrete, which is unusual in osteosarcoma and bone scan did not show uptake in few of the lesions like distal ulnae which were seen on radiographs. This could possibly be due to the small size of the lesion because of recent metastasis

In the literature, the classification Amstutz<sup>4</sup>. for system of multifocal osteosarcoma is the commonly used system. This system divides osteosarcomatosis into types I and II depending on the age group of the patient whether less or more than 18 years in patients with multiple, synchronously occurring lesions within 5 months of initial presentations. Type III comprised those with multiple lesions appearing after 5 months and

subclassified as early metachronous metastatic osteosarcoma (IIIa) when they appeared before 24 months and as late metachronous metastatic osteosarcoma (IIIb) if they appeared after 24 months regardless of age. 1 Another similar classification was suggested by Mahoney which has a similar four-category system of A to D for the above 4 types.<sup>17</sup> These classification systems are of prognostic significance. The synchronous prognosis for multifocal osteosarcoma is poor, with mean survival of six months for type I / Group A and a slightly better range of 5 to 72 months for type II / Group B. Unfortunately, despite advances in both surgery and chemotherapy, more recent reports do not suggest a more favourable prognosis, with a mean survival of 27 months found by Bacci et al, although one patient was disease-free at nine years. 5,16,18,19

Our child would fit into Amstutz type 1 category as the child was eight years old and had multiple lesions at the time of presentation. As mentioned above the prognosis for this category of patients is poor.

This case is reported considering the similarity of the lesions in osteopoikilosis and multicentric osteosarcoma. The osteopoikilosis like lesion may mask the malignant change until late stage. Multifocal osteosarcoma can be misdiagnosed as osteopoikilosis especially when the lesions are asymptomatic. Hence any atypical presentation should be further investigated with a skeletal survey, biochemical investigations, bone scan and biopsy to rule out malignancy, so as to intervene at an early stage.

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#### Announcement

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## SUPRACONDYLAR FRACTURES OF THE HUMERUS IN CHILDREN WHAT IS CONTEMPORARY?

Balakumar.B and Vrisha Madhuri

### New patterns of paediatric Supracondylar humeral fractures

Extension type of supracondylar fractures of the humerus in children are divided into Gartland (modified by Wilkins<sup>1</sup>) type I, II and III. Type I is an undisplaced fracture, type II is a displaced fracture with posterior cortex in contact. Type III is a displaced fracture with no contact. Type III is further subdivided as posteromedial and posterolateral depending upon the direction of displacement. Recently newer types of these fractures have been identified

Leitch<sup>2</sup> et al have described an uncommon subset of supracondylar humeral fractures in nine out of 297 pediatric cases, approximately 3% of their displaced supracondylar fractures. These have instability which is multidirectional and can displace either into flexion or extension as they lack the periosteal hinge. This behavior is confirmed by intraoperative fluoroscopy. They treated the fractures with laterally inserted pin in seven and cross pins in two. Four patients required three pins (all four in the lateral pins group). There was no nonunion, malunion or additional surgery in both the groups. They emphasized the pre placement of K wires into the distal fragment before reducing the fracture and rotation of fluoroscopic arm for lateral imaging as necessary for good reduction. In their group, they were unable to demonstrate a higher degree of associated problems because of the instability. This new fracture which cannot be reduced based on a periosteal hinge was named type IV Gartland fracture.













Bahk<sup>3</sup> et al. have introduced a new sub classification of type III supracondylar fractures based on the fracture lines in coronal and sagittal plane. *Coronal types* have four subtypes which include transverse, lateral oblique, medial oblique and high transverse. *Sagittal types* have a low and high sagittal oblique.

They found out that, greater than 10 degrees coronal obliquity is associated with comminution. Greater than 20 degrees sagittal obliquity was associated with other injuries. The medial oblique fracture could not be held with two lateral pins because due to the fracture obliquity the pins do not hold the medial column well. Similarly high transverse fractures

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cannot be fixed with lateral pins alone. There is a difficulty in classifying this fracture type preoperatively and the classification is done while imaging during the surgery. For management, the recommendation is to get a stable fixation in greater than 10 degree coronal oblique and greater than 20 degree sagittal oblique fractures and high transverse fractures. The problem with greater than 10 degree coronal and greater than 20 degree sagittal obliquity is high chances of malunion because of the following reasons. First is difficulty in obtaining a reduction and difficulty in pinning. Secondly more comminution leads to fixation loss, leading to malunion at a later date. *Third* is poor pin fixation because of obliquity of fracture. High transverse fractures cannot be treated with two or three lateral pins.

Fayssoux<sup>4</sup> et al. have retrospectively analyzed the 422 supracondylar fractures treated by them. They could find 14 cases with fractures involving the metaphyseal and diaphyseal junction defined as the area between horizontal line drawn from the point where the diameter of humerus increased from a constant in diaphysis to increasing value in metaphysis and the horizontal line drawn above the olecranon fossa. Two fracture patterns namely oblique and horizontal were found. Oblique fractures though difficult and time consuming to reduce, are stable after fixation. Horizontal fractures are both difficult to reduce and unstable after reduction. This is because the fracture site is too proximal leading to a long lever arm for reduction. The pin also needs to be inserted with a higher angle which makes catching the opposite cortex difficult. They highlight the identification of such fracture types and suggest a close follow up post operatively with adequate immobilization.

#### Timing of the surgery

Is it essential to do Supracondylar reductions as an emergency?

Classically it is recommended that supracondylar fractures be treated as soon as possible. However in an uncomplicated

supracondylar fracture, where the fracture is partially reduced in the emergency department, the treatment is not compromised by waiting till morning when the OR has orthopedically qualified staff. Mehlman<sup>5</sup> et al performed a double cohort study in children with uncomplicated supracondylar fractures of the humerus. A review of children operated within 8 hours and after eight hours showed that the incidence of open reduction was less in the delayed treatment group

To obtain optimal setting for reduction of uncomplicated supracondylar fractures it is better to wait till next morning rather than do it as emergency in the night with associated problems of a less than optimal environment. Awaiting definitive surgical intervention the next morning, the fracture can be reduced in the emergency department so as to restore partial alignment, correct soft tissue impalement and immobilized in partial flexion. In a study by Mapes and Hennrikus<sup>6</sup> Doppler examination showed decrease flow in brachial artery in positions of increased flexion and pronation.

## Does lack of pulse after a Supracondylar fracture necessitate vascular surgery intervention?

Ghasemzadeh<sup>7</sup> et al showed that capillary refill is a good indicator of whether a vascular surgery consultation is required in a child with absent radial pulse following supracondylar fractures. Of the twenty one patients in their study with pulse-less limbs, only two had delayed capillary refill time. Four received vascular intervention. Of the two with capillary refill time the surgical intervention was not useful because one had a vessel in spasm and the second recovered on its own after fracture reduction and exploration was done before reduction. They suggest vascular surgery consultation only when the absent pulse is associated with delayed capillary refill. An emergency reduction is advocated while awaiting vascular help even in a pulseless pale limb with delayed capillary return.

In a pink pulseless limb reduction of the fracture can be carried out rather than trying to transfer the child to a facility which has vascular surgeons available.

#### Positioning of the child for reduction of Supracondylar fractures

The recommended position for closed reduction is supine with the arm abducted.

Havlas<sup>8</sup> et al have used prone positioning for reduction of supracondylar fractures of humerus and percutaneous pinning. The main advantage of this technique seems to be the ease of operating without an assistant with the gravity aiding reduction. Hyper flexion of the elbow is avoided and hence the chance of ulnar nerve injury is less. The Carm can be rotated to assess lateral view instead of rotating the arm. Insertion of medial wire is easy in this position as extreme external rotation is not needed. This approach is contraindicated in the presence of vascular injury or if other injuries require simultaneous attention. This also may also make the child complications of prone susceptible to anesthesia.

## Does accepting less than anatomical reduction compromise stability?

Minor degree of rotation and displacements are acceptable when reducing a supracondylar fracture. Provided that the carrying angle is maintained, these do not affect the cosmesis or function. However, it does affect the stability of the reduction.

Bloom<sup>9</sup> et al did a biomechanical study on bench top humerus bone model sectioned through the mid olecranon fossa. If the fracture is not reduced completely it is unstable when rotational or varus stress is applied. A third pin either medial or lateral in such situation improves the stability of the low transverse supracondylar fracture.

#### Aiding reduction of supracondylar fracture

Shang Won<sup>10</sup>et al have described the use of a 3 mm K wire inserted from the

posterior aspect for extension Gartland type 3 irreducible fractures that do not regain contact after traction. The pin is passed through the fracture site and is used as a lever to maneuver the distal fragment into place by repetitive flexion and extension.

#### Technique of pinning

Eidelman<sup>11</sup> et al described the use of a 2 lateral and 1 medial pin technique for supracondylar fracture pinning. They found no ulnar nerve injury in 67 patients treated with this technique over a period of 7 years. The lateral pins are placed first with elbow in flexion and the forearm taped to arm with elbow flexed and forearm pronated. The elbow is then extended and the medial wire is inserted and directed laterally from anterior to posterior. Ulnar nerve in this position is less at risk because of a posterior position and is less fixed as cubital tunnel retinaculam is relaxed.

Omid<sup>12</sup> et al in a review of treatment of supracondylar fractures of the humerus have stated that type two Gartland supracondylar fractures are likely to lead to varus malunion and recommended pinning for this injury.

They recommend lateral entry pins widely spaced at the fracture site. It is shown to be as stable as crossed pins in biomechanical and clinical studies. It also avoids iatrogenic ulnar nerve injury.

They feel that angiography is not indicated in a pulseless limb as it delays the fracture reduction and leads to vascular problems.

#### Cubitus Varus. Is it only a cosmetic problem?

Treatment for cubitus varus is often sought for cosmetic consideration.

Gurkan<sup>13</sup> et al have presented three documented cases of posterior instability of the shoulder with associated snapping of medial head of triceps and dislocating ulnar nerve in children following a varus malunion of the supracondylar fractures of the humerus. All three patients had a posterior Bankart lesion on MRI. There was a time interval of several years

between the occurrence of the malunion and development of symptoms at the shoulder. All three children had a significant increased internal rotation. One of the three patients had a voluntary posterior subluxation which also resolved spontaneously after the correction of elbow deformity. The correction of varus and rotation resolved the shoulder symptoms.

In addition to the above study a higher incidence of lateral condyle fracture as a second injury is reported in varus elbows by David<sup>14</sup> et al.

## Cross pins vs. lateral entry pins - an issue of safety vs. stability

Brauer<sup>15</sup> et al in an analysis of 35 studies including 2054 children suggests that the probability of iatrogenic ulnar nerve injury is 5.04 times higher with a medial entry pin as compared to the lateral entry pin. The probability of iatrogenic nerve injury is 1.84 times higher when using medial/lateral entry with isolated than lateral Medial/lateral pin entry provides a more stable configuration, and the probability of deformity or loss of reduction is 0.58 times lower than with isolated lateral pin entry. When the prospective studies alone were analyzed, there were no significant difference in the probability of iatrogenic nerve injury or deformity and displacement, although the confidence intervals were wide. This systematic review indicates that medial/lateral entry pinning, of pediatric supracondylar fractures, remains the most stable configuration and that care needs to be taken regardless of technique to avoid iatrogenic nerve injury and loss of reduction.

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#### **JOURNAL REVIEW**

## SURGICAL DISLOCATION IN THE MANAGEMENT OF PEDIATRIC AND ADOLACENT HIP DEFORMITY

Rebello G, Spencer S, Millis MB, Kim YJ. Clin Orthop Relat Res. 2009 Mar;467(3):724-31 Department of Orthopaedic Surgery, Children's Hospital, 300 Longwood Avenue, Boston, MA 02115, USA.

The authors have retrospectively reviewed their paediatric and adolescent patients who had developmental or acquired hip conditions as a child and underwent correction of proximal femoral deformity using a safe surgical dislocation approach before the age of 18 years. The aim was to assess the efficacy and problems of the surgical approach.

The conditions that were treated were slipped capital femoral epiphysis (SCFE), Perthes disease, developmental dysplasia of the hip, osteonecroses and exostoses. The procedures carried out were mainly femoral head neck osteoplasty (22), intertrochanteric osteotomy (8), and a combination of the two (15). Others were mainly femoral head neck osteotomy, and internal fixation of an acute slipped physis with callus resection. The mean age was 16 years and mean follow-up was 41.6 months. Thirty three of the hips had prior surgical procedure.

In all patients the safe surgical approach described by Ganz was used. The technique allows inspection of femoral head, acetabulum, cartilage and labrum. Any articular cartilage flap and labral tear was debrided and a dynamic assessment of femoro-acetabular contact was performed.

In five patients with unstable SCFE an open reduction using technique described by Leunig et al was performed. That is, after stabilizing the slip and dislocation of the head temporarily, the proximal portion of the stable trochanter was trimmed subperiosteally. The periosteal sleeve of the neck with the blood supply and attached to femoral head was mobilized gently along with the head. The femoral head was reduced anatomically

without tension and fixed with threaded K wires. The postoperative surgical protocol involved limited mobilization and walking bearing one sixth of the body weight.

The Western Ontario Mac Master (WOMAC) score which is a measure of pain, stiffness and function in osteoarthritis of hip or knee was used to assess the outcome. There was a decrease from 9.6 to 5.1 in Perthes and 7.9 to 3.5 in SCFE group. The unstable SCFE had a mean post operative score of 1.2. Seven patients went on to have hip replacement and two had hip arthrodesis.

The major complication was avascular necrosis in 4 patients out of a total of 58 procedures mainly in femoral neck osteotomy (3), and intertrochanteric osteotomy (1).

The technique is technically demanding but the dislocation offered sufficient advantages in assessing and treating these conditions.

#### **Comments**

A number of developmental and acquired abnormalities in children lead to proximal femoral mal-alignment and deformity. causes femoro-acetabular impingement with arthritis in young adult. The management of this impingement due to various causes in the children and adolescents utilizing the safe surgical dislocation technique of Ganz offers advantage of assessment and treatment. However, the technique should be utilized judiciously and after sufficient training. The neck osteotomy for slipped femoral epiphysis in the series had a high complication rate and it is probably safer to pin in the acute stage and then consider osteoplasty combined with intertrochanteric osteotomy for severe deformity.

#### ASSESSMENT OF LOWER LIMB ALIGNMENT: SUPINE FLUOROSCOPY COMPARED WITH A STANDING FULL – LENGTH RADIOGRAPHY

By Sanjeev Sabharwal, MD, and Caixia Zhao ,MD J Bone Joint Surg Am. 2008;90:43-51. Investigation performed at University of Medicine and Dentistry of New Jersey—New Jersey Medical School, Newark, New Jersey

The authors retrospectively compared the measurement of lower limb alignment and limb length discrepancy that were obtained with the use of supine intraoperative fluoroscopy with those that were obtained with the use of full-length standing antero-posterior radiographs of the lower extremity. A fulllength standing antero-posterior radiograph of the entire lower limb was made centered at the knee with the patellae facing anteriorly. The patient placed full and equal weight on both lower extremities and the pelvis was squared with an appropriately sized lift under the shorter lower limb. This method is reproducible and more accurate than clinical methods for assessing frontal plane deformities of the lower limb but cannot be performed intra-operatively. intra-operative fluoroscopic method involved the use of a taut electrocautery cord that is extended from the centre of the femoral head to the centre of the tibial plafond at the ankle followed by an antero-posterior view of the knee. Radiographic analysis involved assessment of the mechanical axis deviation (MAD) and the joint convergence angle (JCA). MAD was calculated as the distance from the centre of the femoral condyles to the vertical line connecting the centre of the femoral head to the centre of the tibial plafond. JCA is measured in degrees as the angle between the distal femoral and the proximal tibial articular surfaces. It was found that the intraoperative fluoroscopy technique with the use of the electrocautery cord method was useful in assessing limb malalignment in patients with a normal body mass index and MAD <2cm and JCA < 3° on the standing radiograph. Age, gender, pelvic height difference and the direction of malalignment did not cause any discrepancy. However the results of fluoroscopy should be interpreted with caution in obese individuals or those who have substantial MAD or pathological laxity of the knee joint. *Comments* 

The fluoroscopic electrocautery cord method is commonly used intra-operatively for assessing lower limb alignment and for planning for surgery on table. The study shows that the technique is not accurate in obese children with large deformities and significant joint laxity, for e.g. in late Blount's disease.

# LATERAL EXTERNAL FIXATION—A NEW SURGICAL TECHNIQUE FOR DISPLACED UNREDUCIBLE SUPRACONDYLAR HUMERAL FRACTURES IN CHILDREN

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J Bone Joint Surg Am. 2008;90: 1690-1697. Investigation performed at the Department of Surgical Pediatrics, Children's Hospital, University of Berne, Berne, Switzerland

The authors describe an external fixation technique using a small monoplanar AO external fixator for complicated Gartland type III supracondylar humeral fractures in children. The basic indications are unsatisfactory closed reduction or lack of stability either due to obliquity of fracture in coronal plane or communition. Open fractures, re-displacements were the other indications. The technique was required in 18.23% (31) of their 170 Gartland type III fractures over a period of 7 years.

An indirect closed reduction could be achieved by this technique in all except one child where there was a soft tissue interposition. Two other children had open reduction because of neurological or vascular injury. The two lateral Schanz pins were inserted perpendicular to the long axis of the fragment; the proximal one 2 cm above the highest fracture line and the distal through the centre of the capitellum and were used as

joysticks for reducing the fracture. The fixation was augmented by a derotation 1.6 or 2 mm K wire inserted from lateral aspect ante or retrograde crossing the fracture just above the olecraon fossa. Schanz pins were connected to a rod. Additional immobilization in the nature of a dorsal splint for comfort for few days was provided for some children. Mean time to fixator removal was 5 weeks. Mean follow-up was 41 months

The outcome in terms of limb alignment was excellent, the elbow range of motion was restricted in a single case and the patient satisfaction was high in all cases. There was no growth arrest. The pin track infection in this series was insignificant(2) and easily treated with oral antibiotics

#### **Comments**

The method appears to be a safe alternative in cases where closed reduction is not attained or sufficient stability is not achieved with standard methods of closed reduction and Kirschner wire fixation. The pin track infection could be expected to be more significant in India. The duration of immobilisation in this study is higher than normal for this fracture in children. In this study all cases were supervised by the senior author (TS). There could be greater risk of growth plate injury in the hand of inexperienced surgeons as the diameter of the screws are large and multiple attempts in the distal fragment could damage the physis The major advantage of this technique is in achieving an indirect closed reduction in irreducible fracture avoiding thus avoiding open reduction and providing sufficient stability to not need cast immobilization. It is a useful alternative to open reduction and in open fractures and vascular injuries.

## FAILURE OF ORTHOFIX EIGHT-PLATE FOR TREATMENT OF BLOUNT DISEASE

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J Pediatr Orthop. 2009 Jan/Feb; 23 (1): 7-60 The Department of Orthopaedics, Medical College of Georgia, Augusta, GA.

Surgical treatment options for Blount disease and other angular deformities around the knee include hemiepiphysiodesis and corrective osteotomy. Hemiepiphysiodesis has gained acceptance as a minimally invasive technique to correct deformity gradually. A number of hemiepiphysiodesis techniques have been used, including direct ablation of one side of the physis, transphyseal screw placement, and staples. Recently, the eight-Plate (Orthofix, McKinney, Tex), a tension band device, for directed growth has been introduced with good results.

This study analyses the results of guided growth by use of eight plates to correct the angular deformity of the knee in 24 children with 31 angular deformities. The emphasis is on understanding the failures and complications of the guided growth by the 8 plate device.

Of the 31 extremities, all received proximal tibia plates, and 10 also received distal femur plates. The deformities were due to Blount disease, genu varum and genu valgum. Mean length of follow-up was 17 months for all patients with a minimum of 1 year. The surgery was performed on an outpatient basis, inserting 1 plate per physis submuscularly while preserving the periosteum. Ambulation was not restricted.

Implant failure occurred in 8 (26%) of 31 constructs. All 8 failures occurred in patients with Blount disease and involved breakage of the tibial implant's metaphyseal screw. The mean time to failure was 13.6 months. Eight implant failures in 18 extremities within the Blount group represent a failure rate of 44%. No implant failures occurred in those with other diagnosis. No distal femur implants experienced screw or plate failure. Within the Blount group, none progressed after hemiepiphysiodesis. Of the 10 patients who did not experience hardware failure, 5 were corrected fully, requiring hardware removal. Four were corrected partially, averaging 8 degrees of

correction, and 1 had no correction. Mean correction for genu varum and genu valgum was approximately 5 degrees/year .

The eight-Plate (Orthofix) is temporary, and its extraperiosteal placement allows for easy removal without physeal damage and a lateral fulcrum creating a longer moment arm for the remainder of the physis to grow and correct the angular deformity. This reduces the dependence on the imprecise science of estimating the remaining growth and allows children of all ages to be candidates for surgery.

It may be a combination of the excess weight and the physeal anatomy in patients with Blount disease that contributes to the screw breakage. The cyclic loading of the varus knee with every step along with an abnormal pathologic condition could create significant motion at the physis and cause the screws to fail secondary to fatigue. Accordingly, no distal femur implants failed even in the heavy patients with Blount disease, possibly because the physis within the distal femur even in Blount disease is still normal.

Interestingly, the hardware that failed was consistently the distal/metaphyseal screw. The screws used in the eight-Plate (Orthofix) are 4.5-mm cannulated titanium screws.

Currently, the eight-Plate (Orthofix) system offers no noncannulated screws or screws with a larger core diameter. Cannulated screws have similar pullout strength to cortical screws but have 16% to 27% less strength in torsion and 3-point bending. Core diameter is the principal factor in determining fatigue life. Titanium screws are significantly weaker than steel screws of the same core diameter. Noncannulated screws of the same core diameter would be stronger. The cannulated feature is unnecessary for this straightforward procedure.

#### **Comments**

The authors outline the benefits of" temporary" hemiepiphysiodesis over all other methods of angular deformity correction about the knee and describe their experience with the Eight

plate. They report the failures in their series and stress the fact that all the failures belong to the Blounts group which might be due to the high BMI and the pathologically mobile abnormal physis. They suggest using noncannulated stainless steel screws with large diameter, which will make the system more failure proof.

# POST CLOSED REDUCTION PERFUSION MAGNETIC RESONANCE IMAGING AS A PREDICTOR OF AVASCULAR NECROSIS IN DEVELOPMENTAL HIP DYSPLASIA

Carl Tiderius, Diego Jaramillo, Susan Conolly, Kasser, Michael Griffey, Diana Rodriguez, Michael Millis, James .R, David Zurakowski, Young-Jo Kim. Dept of Orthopaedic Surgery and Radiology, Children's Hospital Boston, Children's Hospital Philadelphia. J Pediatr Orthop 2009;29:14-20.

The authors looked at the predictive ability of post closed reduction contrast enhanced MRI for avascular necrosis (AVN) after closed reduction in DDH. The study was a retrospective analysis. Twenty eight hips in twenty seven infants (mean age 5.5 months, range 1-11 months) with idiopathic hip dislocations which had failed brace treatment, underwent surgical treatment of adductor tenotomy and closed reduction were included in the study. Pre operative traction was not used. Cast was changed on a monthly basis, time averaging around three months, followed by abduction bracing.

Following closed reduction and casting (the first procedure) confirmation of reduction was obtained with MRI within 24 hours. MRI scan was usually performed while still recovering from anesthesia. In a rare case sedation was used. Gadolinium was given IV and MRI was performed where the filling of the vessels and the physis, vascular canals and finally diffusion into epiphyseal cartilage was studied. The children were followed by serial radiographs until a minimum of 13 months after reduction (mean 22 months; range 13-42 months). They were studied to assess the

appearance of AVN. The children were clinically followed up to 12 years (2-12).

Six of 28 hips (21%) developed radiological evidence of AVN. Fifty percent of the hips with AVN but only two out of 22 without AVN showed global decreased enhancement (p<0.05%). A global decreased enhancement in the MRI irrespective of the abduction angle and age, had a significantly higher risk of developing AVN (p<0.001) independent of age at reduction (p=0.02) and abduction angle.

There was a trend towards older age group (7.3 months), at reduction, in the AVN group. The angle of abduction <55° or > 55° between the groups however showed no correlation to the development of AVN. The authors concluded that the perfusion MRI may be the first investigative tool capable of predicting future AVN at a time when ischemia is still potentially reversible. A major limitation of the study was the limited specificity of global decreased perfusion enhancement in predicting AVN because even affected hips exhibited normal radiological development in 50% of the cases similarly 50% of the cases which did not show a decrease developed AVN. They advocated MRI soon after reduction was over as there are only porcine studies which showed reversibility of ischemia within 6 hours and no data exists beyond that time. This study opens a door to future studies looking at changes in hip position and reduction methods that may potentially decrease the AVN rates.

#### Comments:

This is the first human study to fashion a clinical tool to predict the possibility of AVN occurring after a closed reduction and spica casting. Clinical use of this technique in India may be dictated more by the availability on site. It was interesting to note that the safe limits for abduction considered to be less than 55° in this study did not influence the AVN and it appears that limiting abduction alone may not prevent AVN. The abduction angles in this study however are on the higher side (mean 58°),

range 45-75) both for the AVN and (mean  $55^{\circ}$ , range 40-70) for the non AVN group.

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#### Diagnostic Quiz

A 13 year old boy presented with progressively increasing swelling of posterior aspect of right thigh and knee of 2 years duration. There is a past history of excision of a swelling over the trochanteric regions. Four other siblings are normal.

Redness and swelling of the right elbow was noticed at the time of planning for surgery on this patient. There was also firm swelling over the extensor aspect of left elbow

Investigations:
ESR 108mm/ hour
WBC counts within normal limits
Hb.- 11.6
Rheumatoid factor negative
LE cell negative
Serum creatinine-0.4 mg%
Serum calcium-8.8 mg%
Serum phosphorus-5.8 mg%
Clinical photograph and radiographs

