CONGENITAL UPPER LIMB DEFORMITIES

SWANSON'S CLASSIFICATION

1. Failure of Part formation

Transverse absence:	Congenital amputation
Longitudinal absence:	Radial Club hand
	Ulnar Club hand
	Cleft hand

2. Failure of Part Differentiation

Radioulnar synostosis
Camptodactyly
Trigger thumb
Syndactyly

3. Duplication Polydactyly: Pre-axial, Post-axial

4. Overgrowth Macrodactyly

5. Undergrowth Thumb hypoplasia (Blauth)

6. Miscellaneous

Constriction ring syndrome
Congenital dislocation of radial head
Madelung's deformity

FACTS

Congenital anomalies affect 1% to 2% of newborns
Approximately 10% of those children have upper-extremity
Genetics

Trisomy of 21, 18, 13	Polydactyly, Syndactyly
Apert's syndrome	AD

Environmental factors:

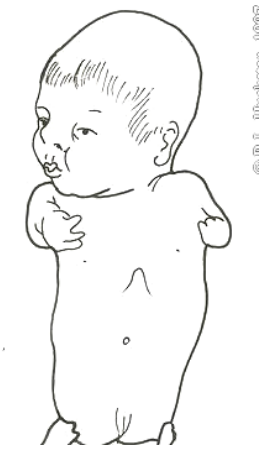
Drugs: Talidomide, Dilantin, Warfarin

ASSESSMENT

Initial visit: an emotion filled event.
Parents: Underlying guilt.
Parents have difficult for families to accept.
Risk for Future Pregnancies: Genetic counselling

TRANSVERSE AMPUTATION

Part distal is missing.
Rare
Eg: Absence hand at wrist



INTERCALARY AMPUTATION

Segment in between is absent.
More common with Talidomide Intake during pregnancy

RADIAL CLUB HAND

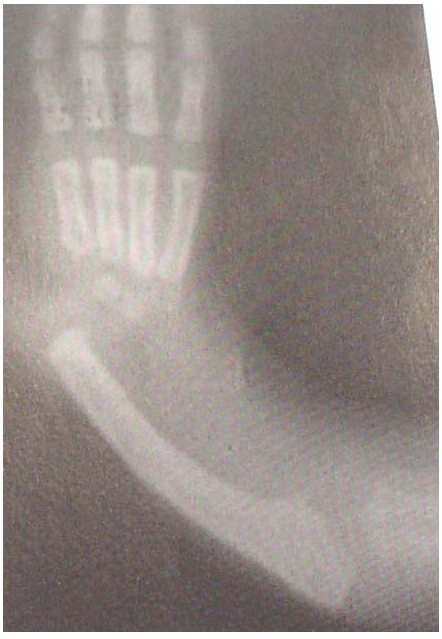
1: 30,000

Bilateral in 60%

Clinical

Absence of radius and radial rays
Bowing of the ulna

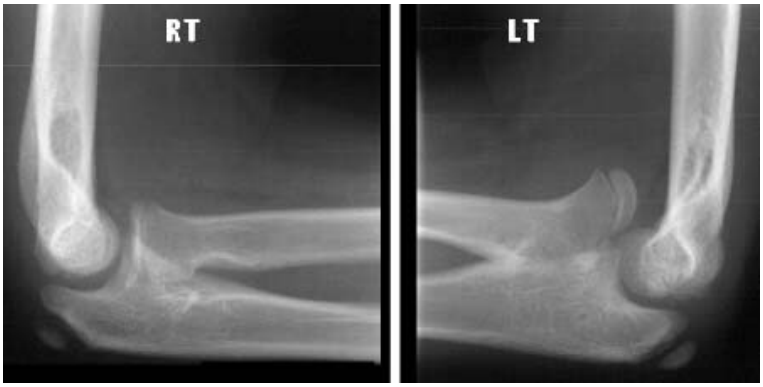
Associated
- Anal atresia
- Tracheo- oesopharyngeal fistula
- Radial bone absence
- Renal anomalies
- Cardiac anomalies



Treatment

Use of serial casts
Realignment and stabilization: Lower end of ulna is transferred radially.
Tendon transfers - Ulnar side tendons transferred radial side
Correction of the ulnar bowing – osteotomy

RADIO-ULNAR SYNOSTOSES



AD;

Bilateral in 60%

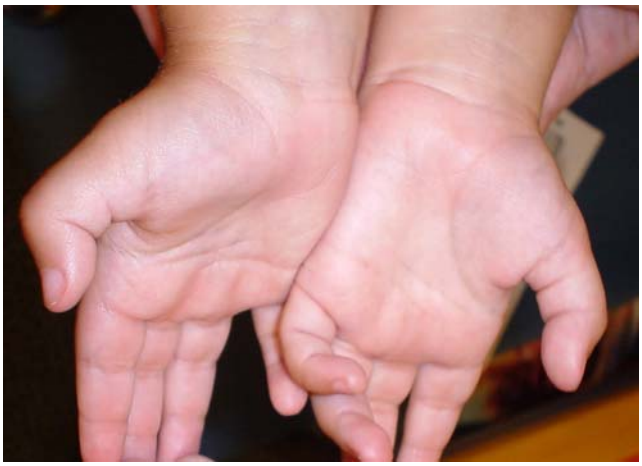
Site: Proximal 1/3 of forearm

Deformity: Forearm pronation

Rx: Leave it alone.

When bilateral: rotational osteotomy in one forearm to a functional position

TRIGGER THUMB



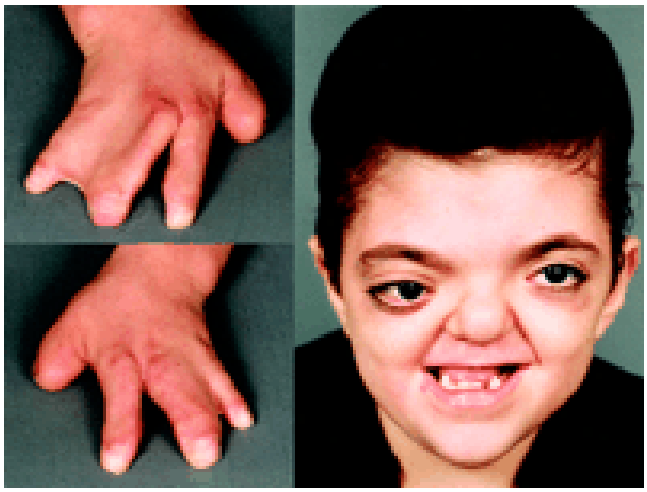
Often bilateral, with fixed flexion of thumb

If present >1 yrs Rarely recover

Treatment: - Surgery

Release A1 pulley [base of the tendon sheath]

SYNDACTYLY



Syndactyly: Syn = together in Greek

Mnemonic 5,15,50,30:[Thumb and Index 5%;
Ring and little: 30%]

More common in white males, AD

Present bilaterally in 50%

Types: I Isolated

II Complex Trisomy 13, 14, 21

Apart syndrome [clinodactyly; facies]

Holt Oram Syndrome [ASD, Radial club hand]

Poland's [Absence of Pectoralis major]

Treatment

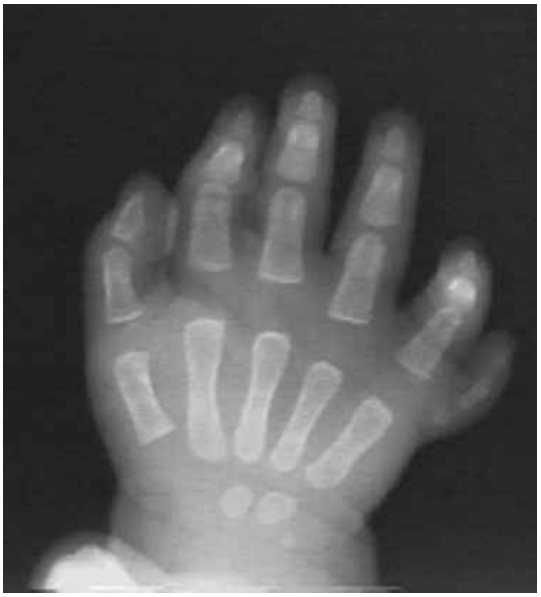
Surgery: At 18 months old.

Full-thickness skin grafting is almost always required for soft tissue coverage.

Technique

Mark the skin flaps Z plasty
Dorsal butterfly flap, to make the web
Both sides of the same digit : never separated on the same day (>6 wks)

POLYDACTYLY



POST AXIAL POLYDACTYLY

Post axial [next to the little finger]

Is a common hand anomaly

10 times more in Blacks

AD

Nonsyndromal

PRE AXIAL POLYDACTYLY

Preaxial [next to the thumb]

Is common duplication in whites

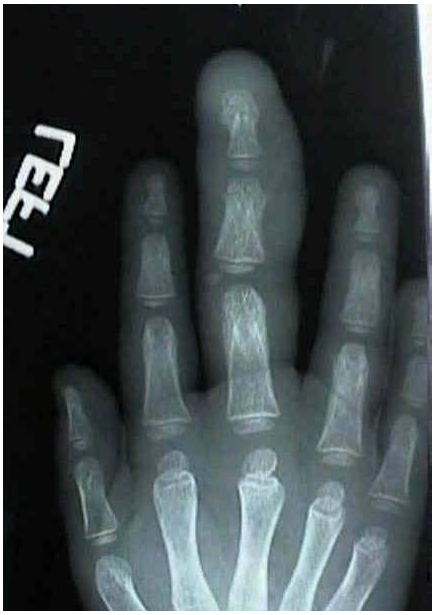
AR [17 chromosome]

Syndromal

Treatment

Excise the rudimentary finger

MACRODACTYLY



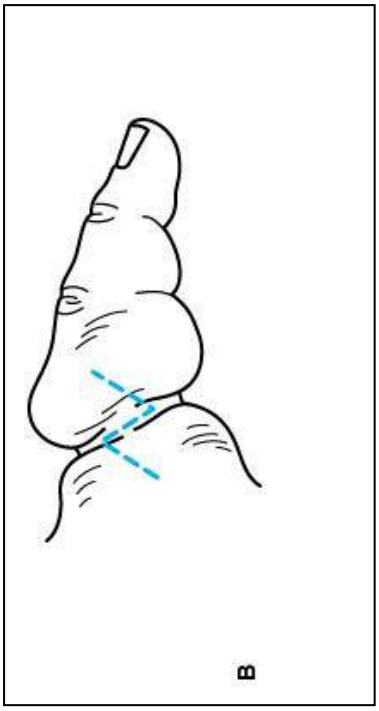
-Hamartomatous enlargement of soft tissue

-Associated with neurofibromatosis

When hemi-hypertrophy, suspect
Wilm's tumor
Adrenal carcinoma
Hepatoblastoma

Treatment Debulking,
Osteotomy
Epiphysiodesis

CONSTRICTION RING SYNDROME [CRS]



Also called: Streeter's syndrome

1 in 15,000 live births

The etiology of CRS is unclear

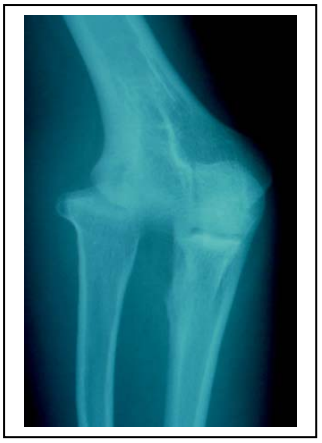
It is not genetic

CRS frequently affects both arms and all four limbs

Commonly associated with clubfoot

Surgical treatment: Z-plasty of constriction

CONGENITAL DISLOCATION OF THE ELBOW



Incidence: 0.16%

D/D

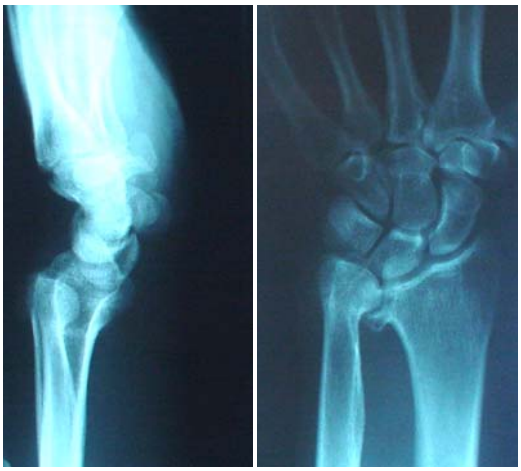
Old Monteggia
Multiple endochondromatosis or exostosis
Radio-ulnar synostosis

Clinical: Family history; Bilateral anomaly;
Loss of supination

Radiological: Hypoplasia of the Capitullum
Dome shaped Head

T**reatment:** If painful, excision after maturity

MADELUNG'S SYNDROME



Epiphyseal arrest on ulnar and volar half of the distal radius

Joint is directed ulnarward and volarward

AD variable expressivity; Females (4:1)

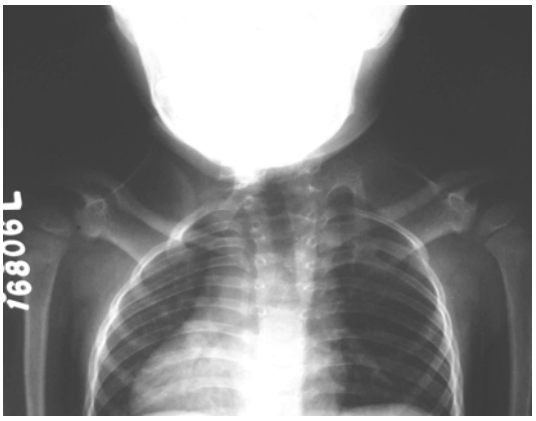
Wrist motion: Extension and supination are limited

X ray may show: Classical deformity
Inferior Radio-ulnar dislocation

Treatment Corrective osteotomy

Physeal resection and fat graft`

SPRENGEL'S SYNDROME



Failure of the normal caudal migration of the scapula

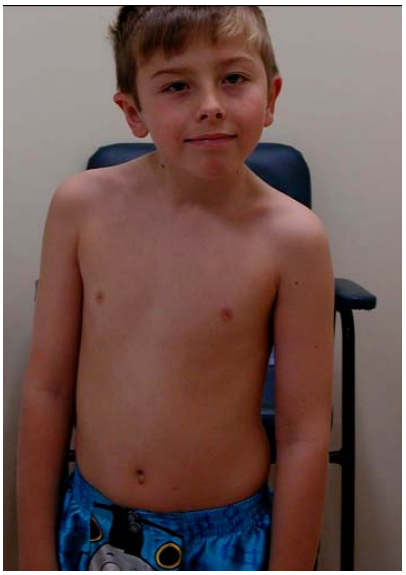
Associated with Congenital scoliosis

Klippel Fail syndrome

The superomedial scapula is connected to the proximal cervical spinous process through an omovertebral connection (bone, cartilage) in 30%

Treatment: Surgical correction

TORTICOLLES



Associated with other “Molding disorders”

Hip dysplasia
Metatarsus adductus
Plagiocephaly

Treatment Stretching exercise 90% good results

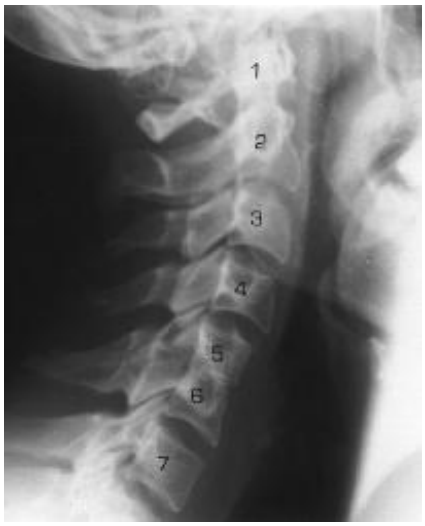
. Surgery if necessary at one year (Distal or proximal release or both)

Look for Upper cranial anomaly

Ophthalmologic disorder

Posterior fossa brain tumor

KLIPPEL FAIL SYNDROME



Low hair line

Limited ROM of the neck

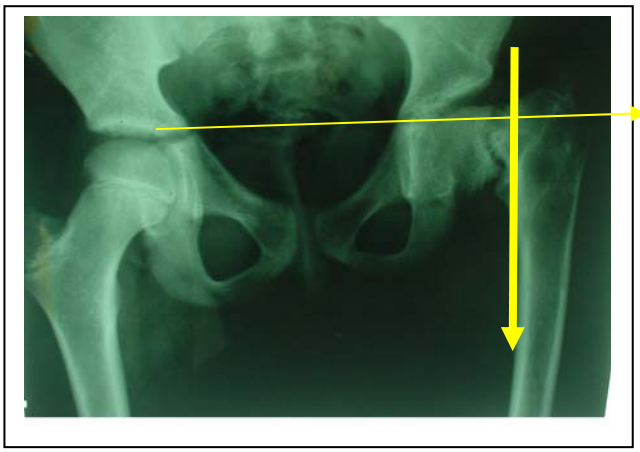
Can be associated with Sprengle’s shoulder,

Renal or cardiac anomaly

Avoid contact sports

3. LOWER LIMB CONGENITAL DEFORMITIES

INFANTILE COXA VARA



Clinical

Trendlenburg gait
Adduction is more than abduction
Supratrochanteric shortening

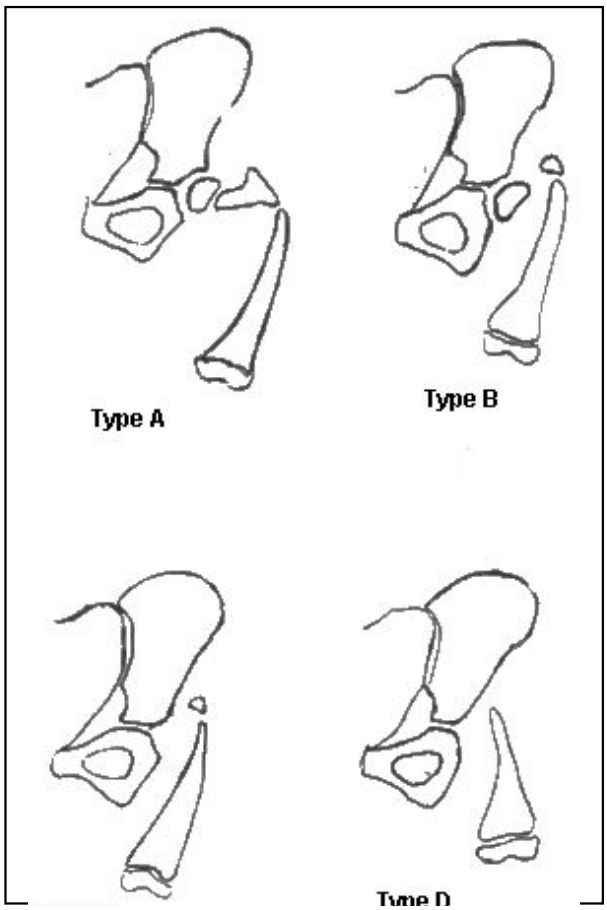
X ray

Coxa Vara <110º

Hilgenreiner epiphyseal angle > 60 º

Treatment Corrective Valgus osteotomy

PROXIMAL FEMORAL DEFICIENCY



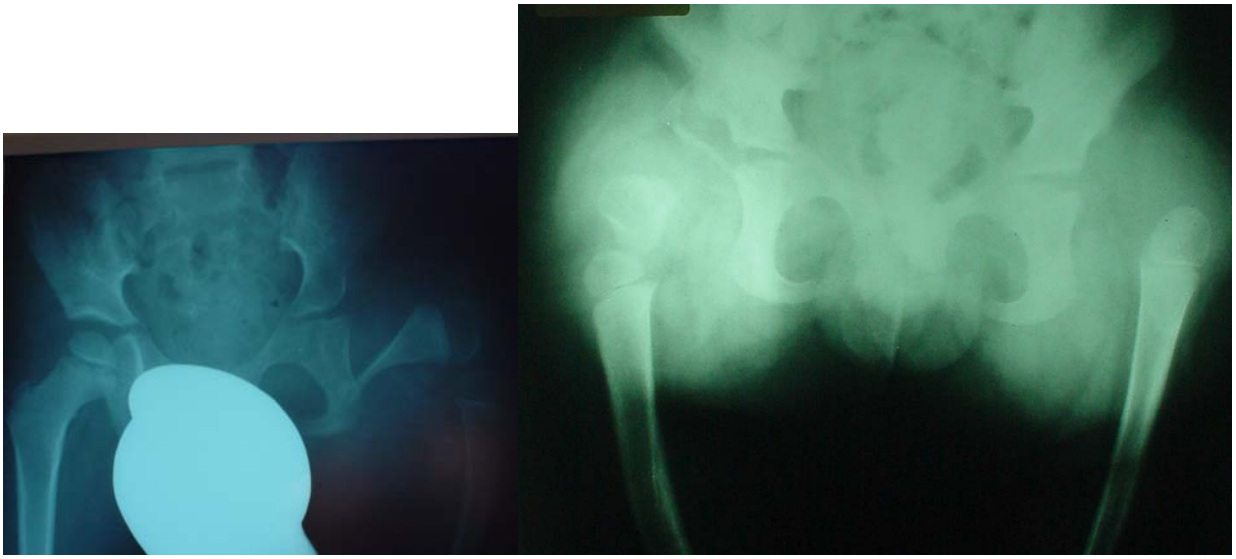
Can be associated with absence of fibula.
tarsal coalition is common

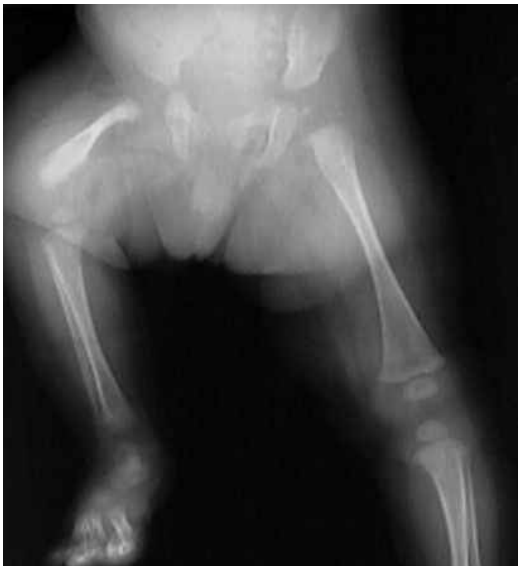
Type A [congenital short femur]
Gap between proximal and distal femur, which ossifies by maturity
Femur is 80% short

Type B
The femoral head and acetabulum are hypoplastic;
The femoral gap does not unite

Type C
The Femoral head is absent
The Acetabulum is present but dysplastic

Type D
The femoral head and acetabulum: absent





Treatment

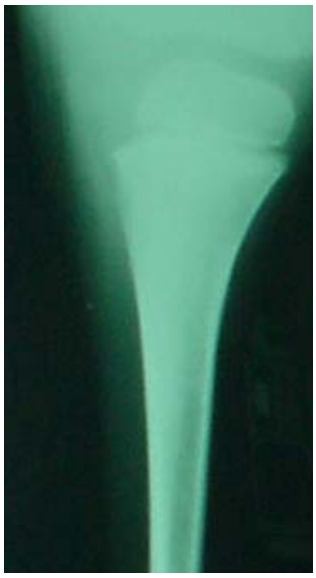
Type I Ilizarov lengthening of the femur

Type II Valgus osteotomy and fusion

Type III and IV

Extended prosthesis
Syme's and extended prosthesis
Knee fusion and Syme's [Brown]
Van Ness procedure: Informed consent and Psych evaluation

ABSENCE OF FIBULA



Absence of Fibula more common than tibia

Can be associated

PFFD,
ACL absence,
Toe ray absence

Tarsal coalition

Tibial bowing: Anteromedial bow

Treatment Syme's amputation

POSTEROMEDIAL BOWING TIBIA

Postural

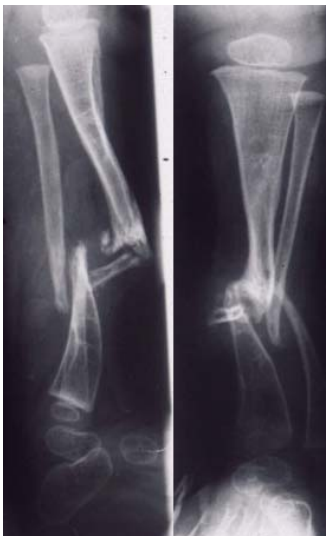
Posteromedial bowing of tibia

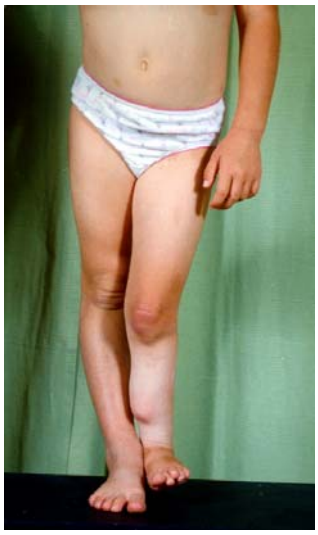
Is usually benign

Observe for shortening of the limb

Treatment: Regresses with stretching exercises

ANTEROLATERAL BOWING OF TIBIA





Rare but serious condition

Treatment

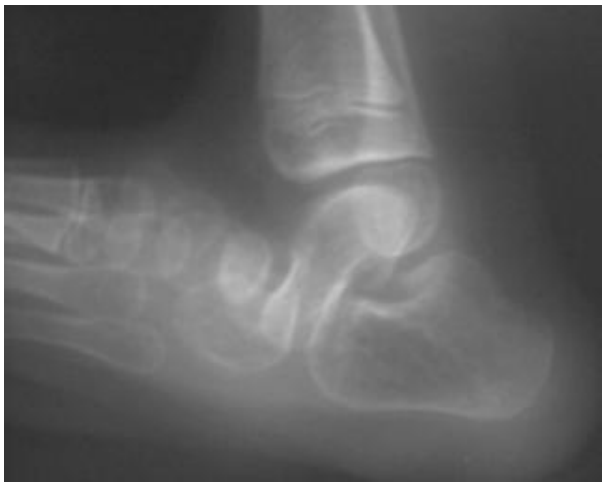
Protective brace

Bone graft and fixation of tibia

Some times repeat operation

May need disarticulation of the ankle

CONGENITAL VERTICAL TALUS



Autosomal dominant

Associated with: trisomy of 13

Talus remain in plantar flexion both in dorsiflexion and plantarflexion.

Treatment: Soft release and plaster cast