

# **SICKLE CELL ANEMIA**



Normal cell



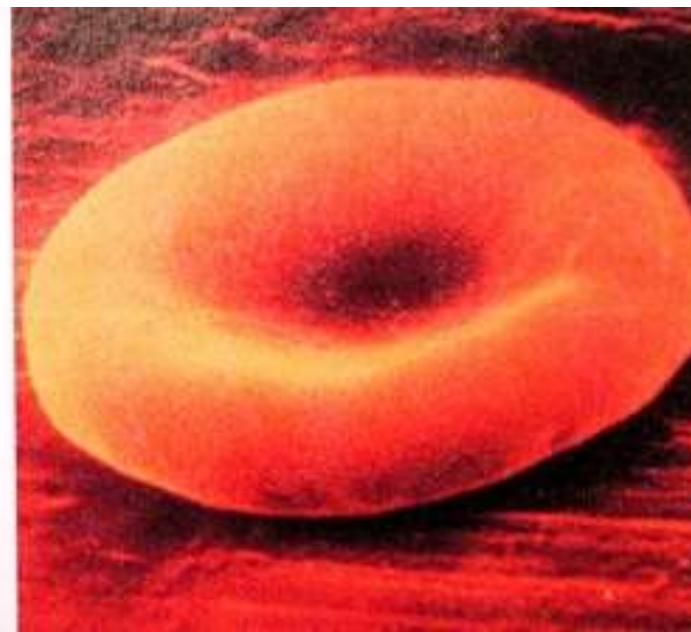
Spherical cell



Oval cell



Sickle cell



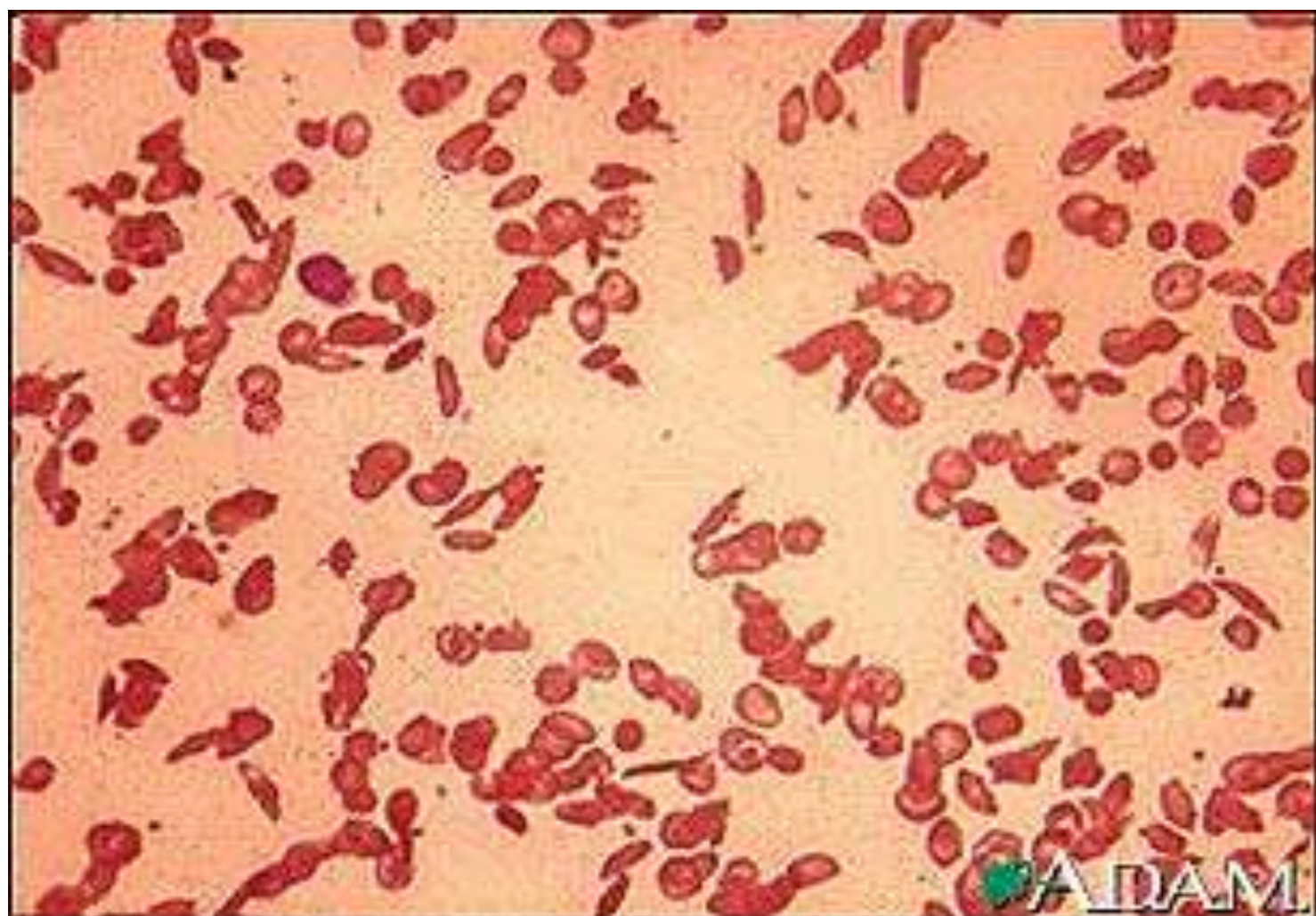


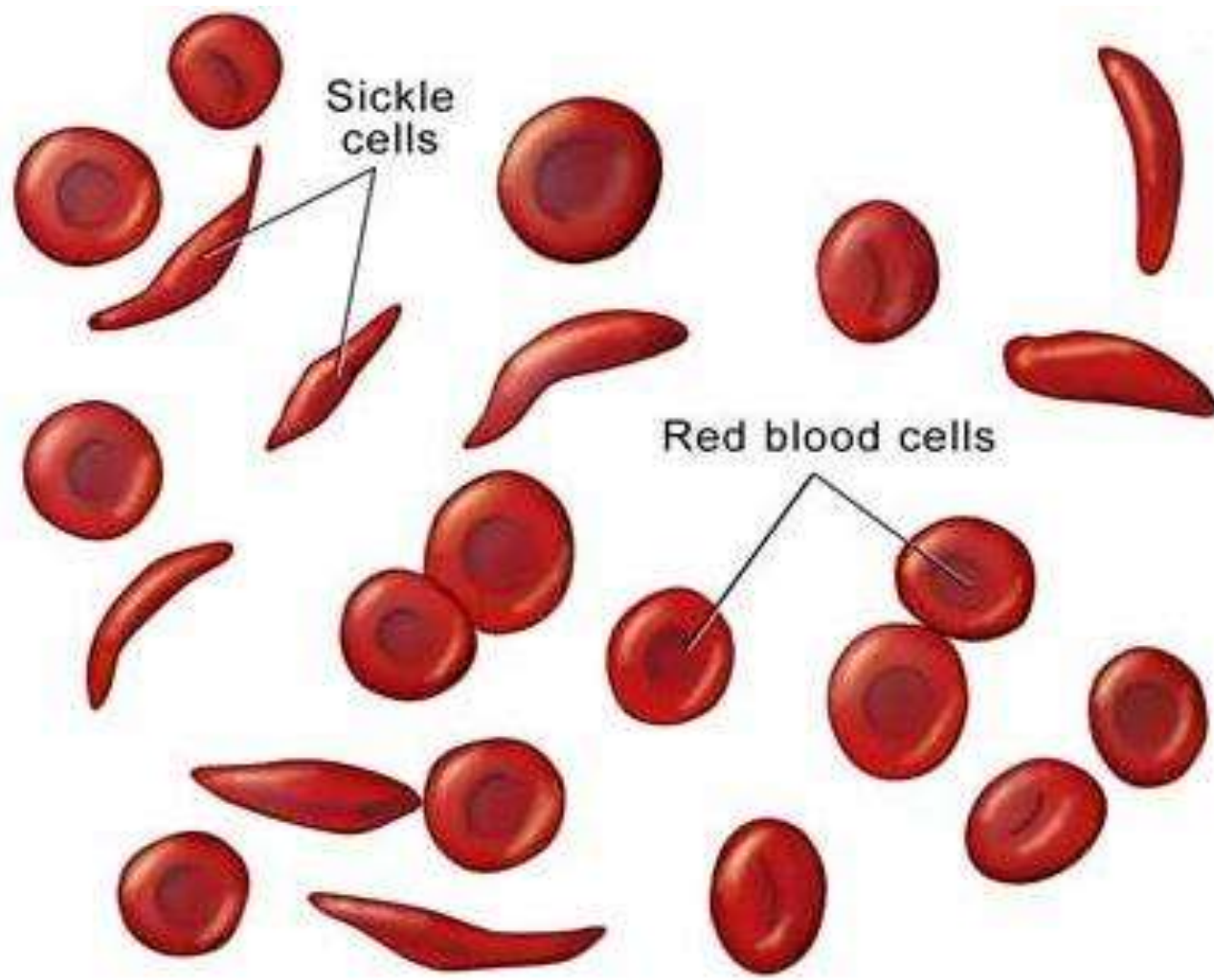
Normal red blood cell



Sickled red blood cell







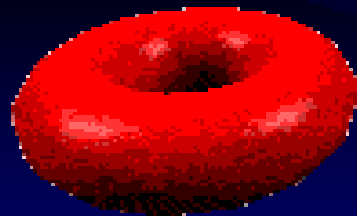
Sickle cells

Red blood cells

# Normal vs. Sickle Hemoglobin

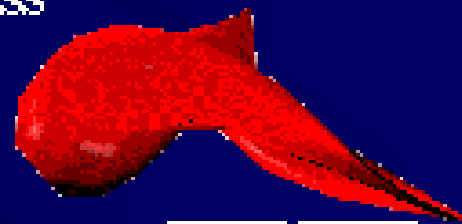
## Normal

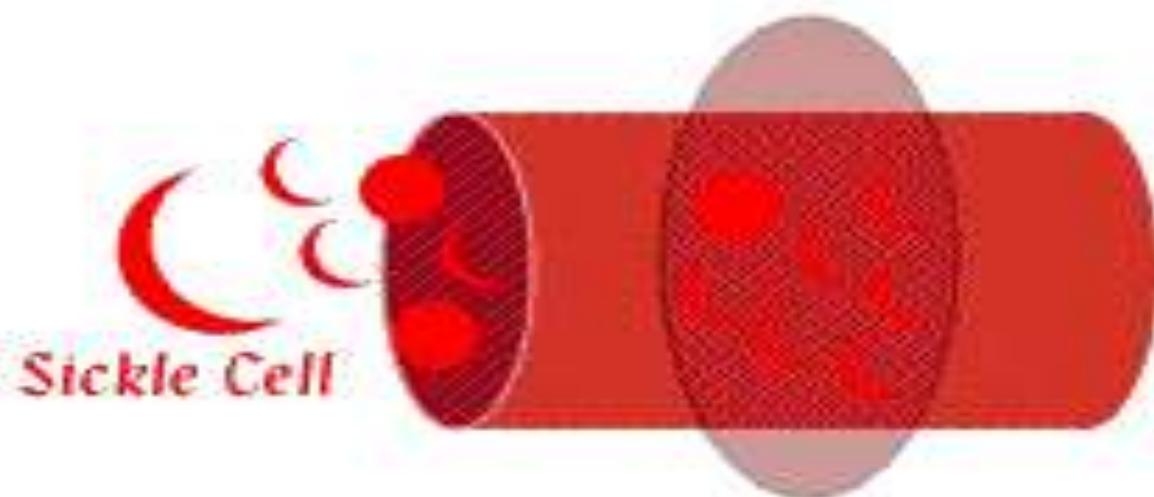
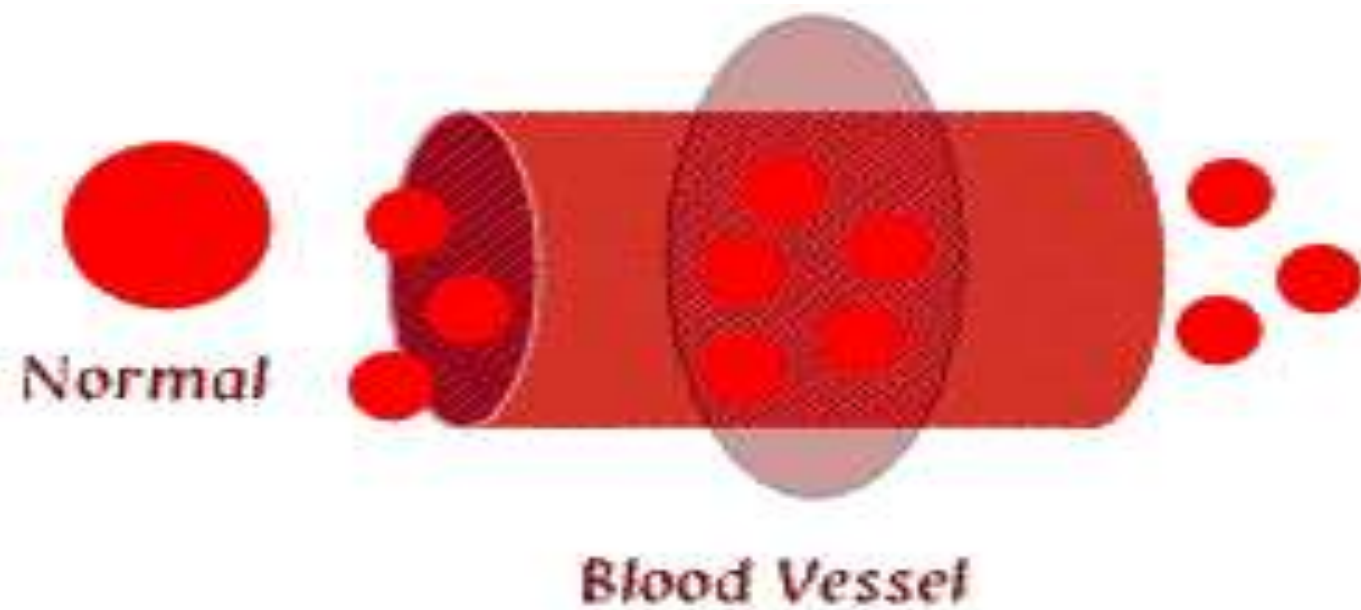
- disc-Shaped
- soft (like a bag of jelly)
- easily flow through small blood vessels
- lives for 120 days



## Sickle

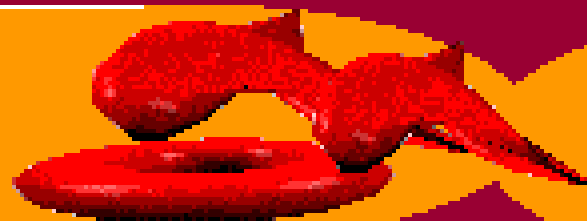
- sickle-Shaped
- hard (like a piece of wood)
- often get stuck in small blood vessels
- lives for 20 days or less



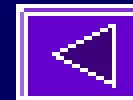




# Normal vs. Sickle red cells



If no oxygen, then pain  
and damage occurs





Normal hemoglobin



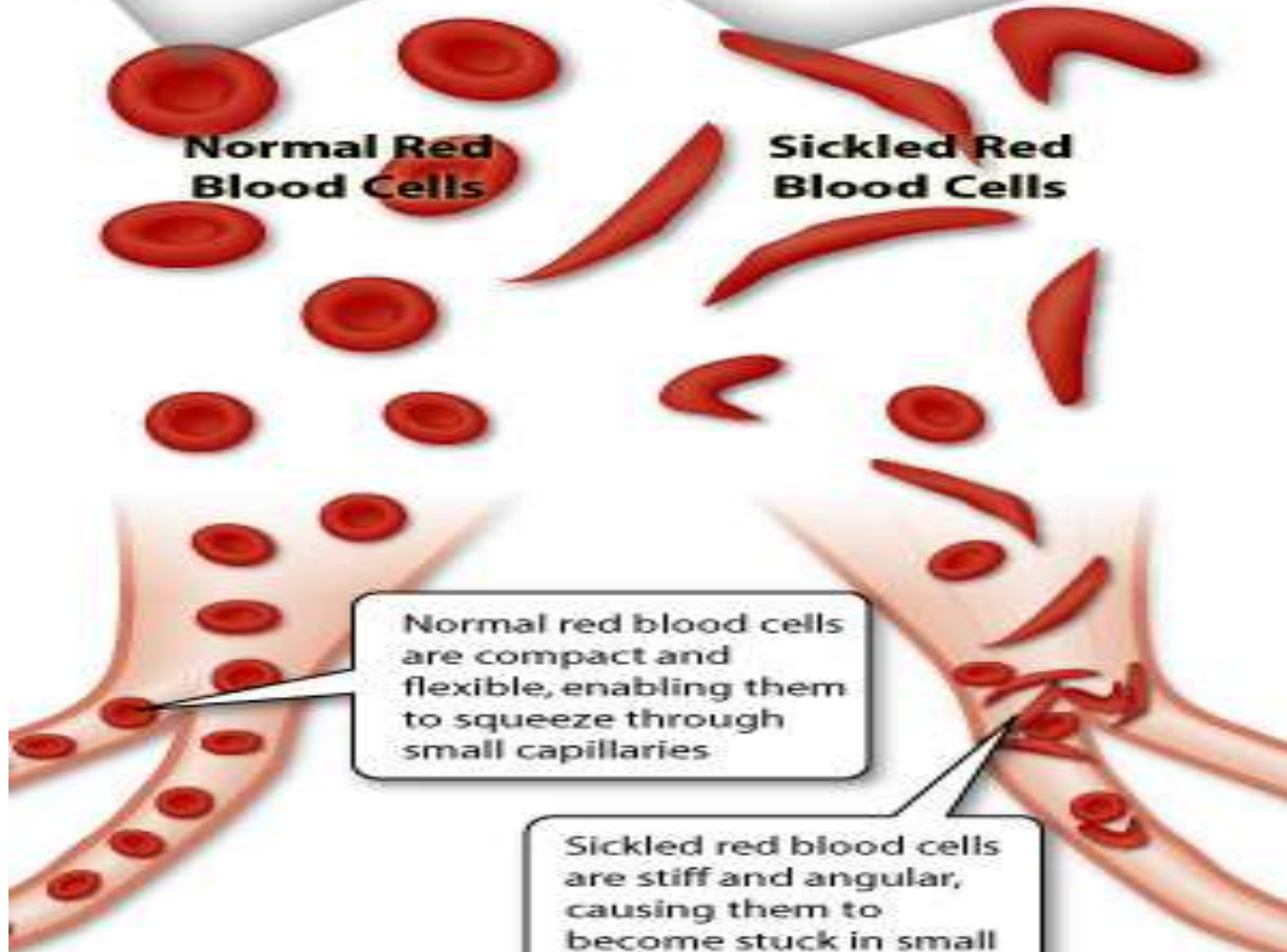
Sickle Cell hemoglobin forms long, inflexible chains

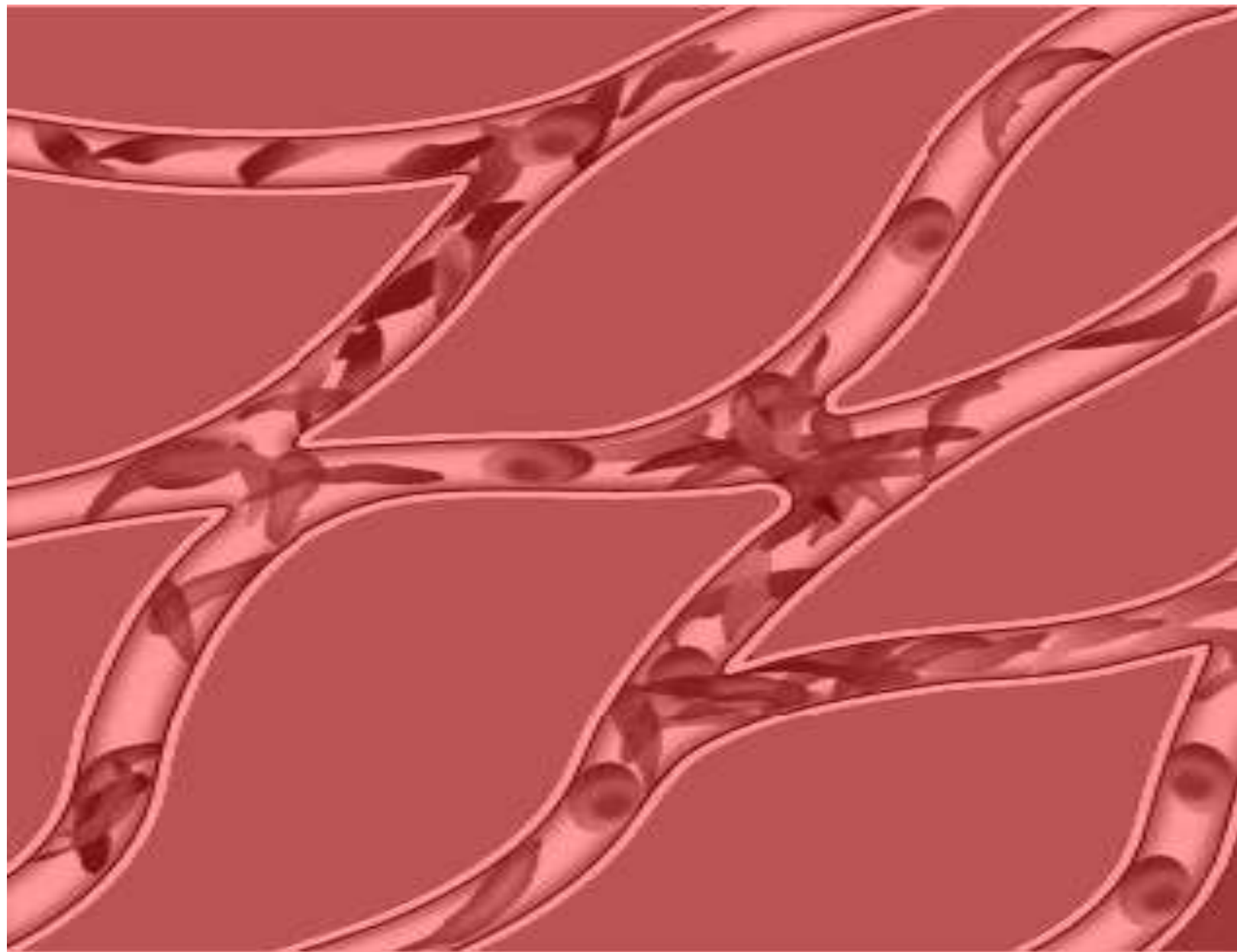
**Normal Red Blood Cells**

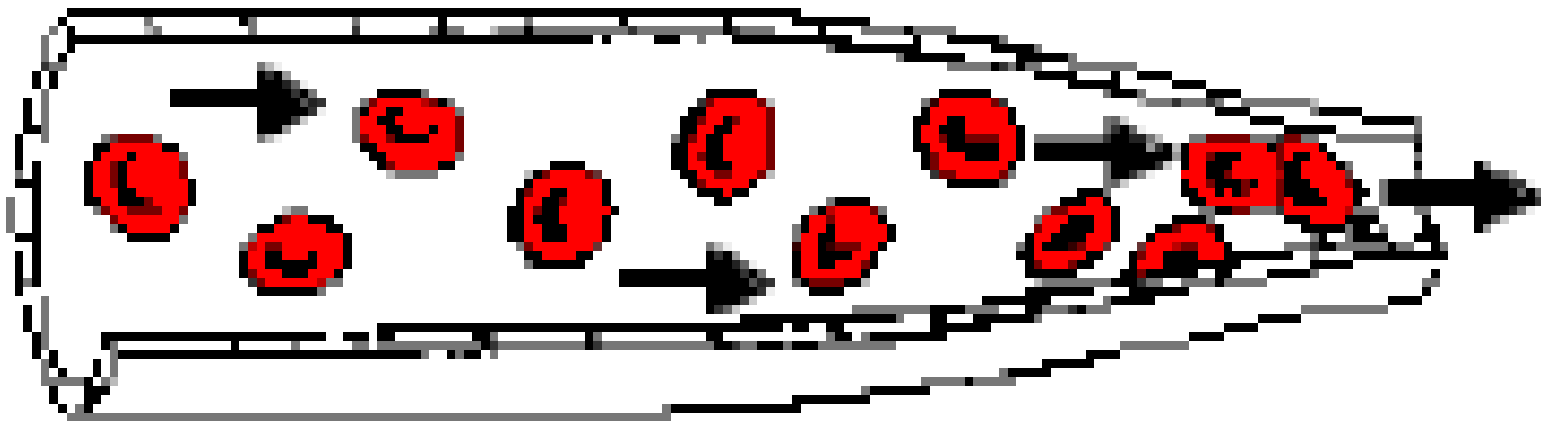
**Sickled Red Blood Cells**

Normal red blood cells are compact and flexible, enabling them to squeeze through small capillaries

Sickled red blood cells are stiff and angular, causing them to become stuck in small capillaries





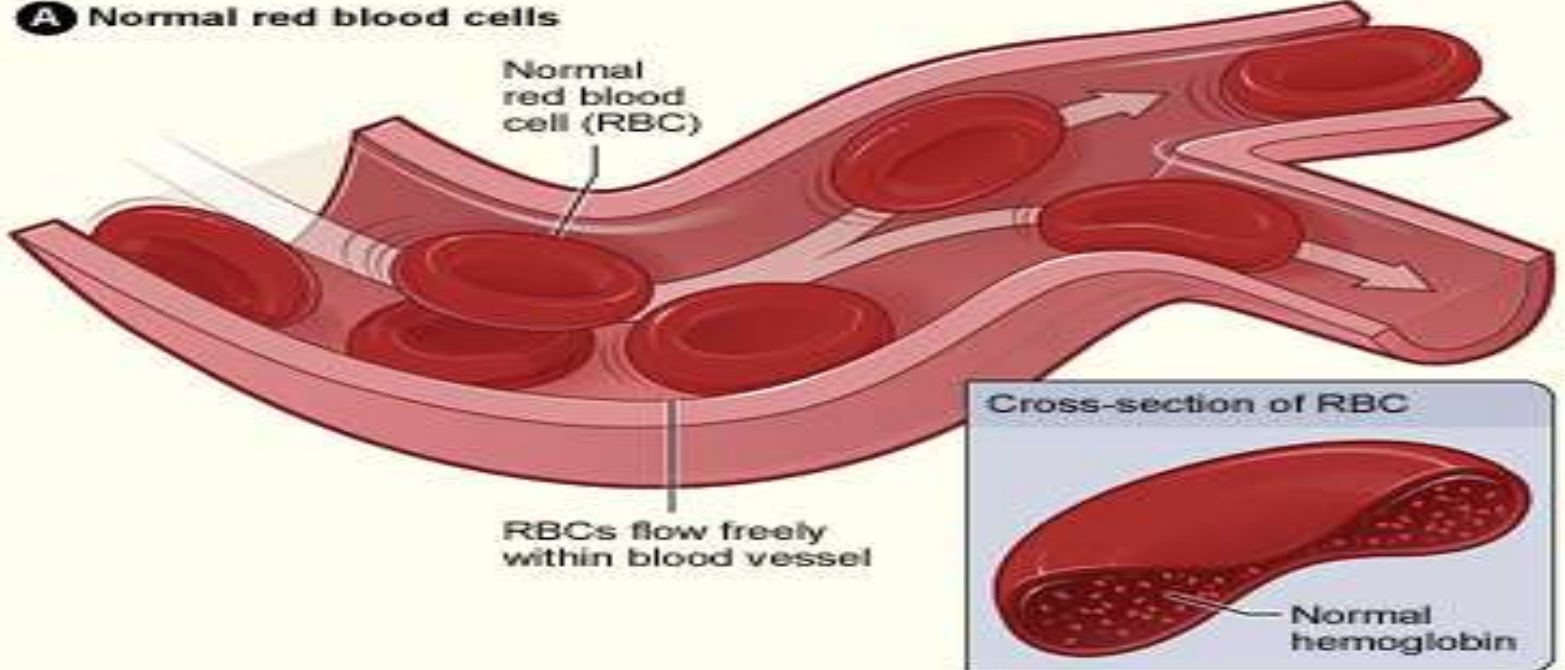


Normal cells move freely

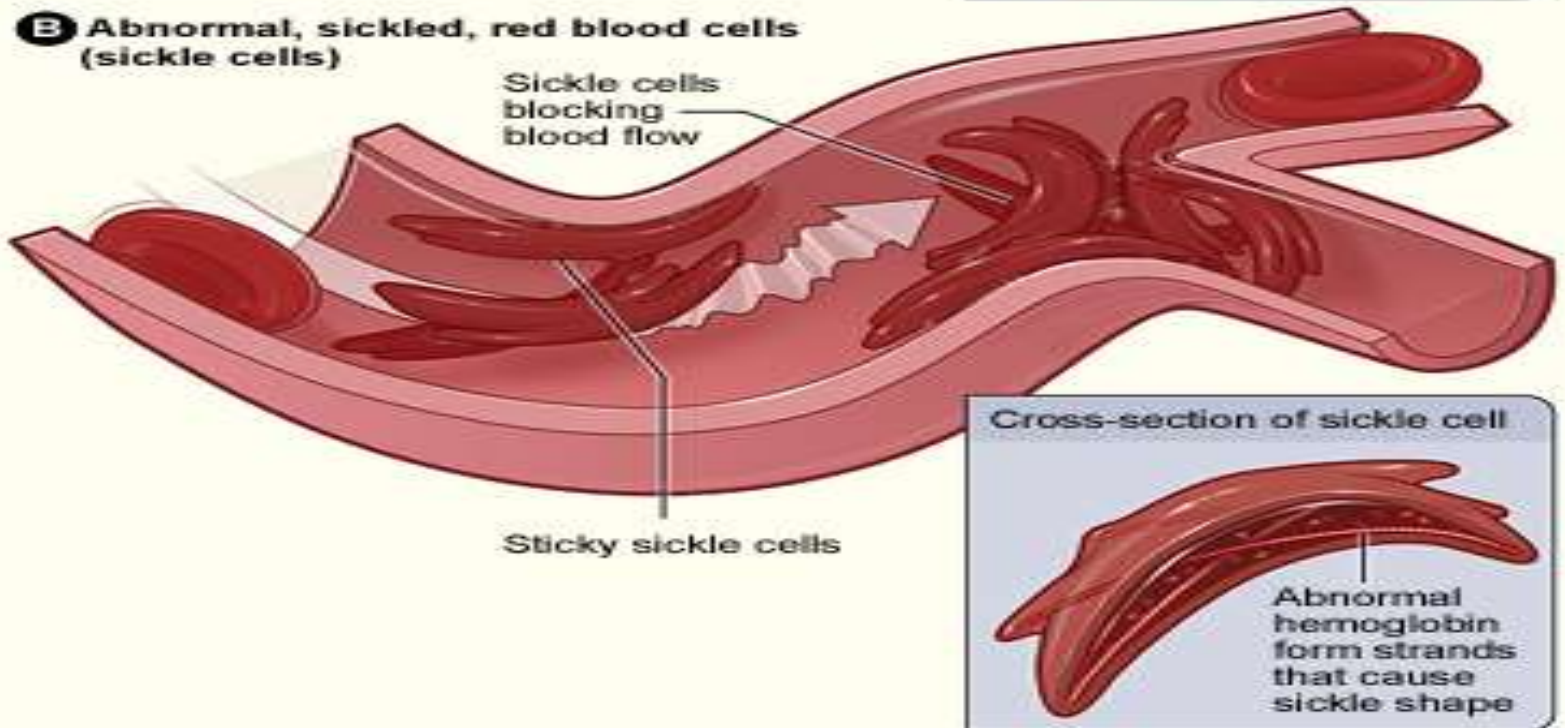


Sickled cells get stuck

**A Normal red blood cells**



**B Abnormal, sickled, red blood cells (sickle cells)**



## INTRODUCTION

- First described in Chicago in 1910 by James Herrick as an inherited condition that results in a decrease in the ability of **red blood cells** to carry oxygen throughout the body
  - Sickle red blood cells become hard and irregularly shaped (resembling a sickle)
  - Become clogged in the small blood vessels and therefore do not deliver oxygen to the tissues.
  - **Lack of tissue oxygenation** can cause excruciating pain, damage to body organs and even death.

# DEFINITION

- Sickle cell hemoglobinopathies are hereditary, genetically determined (autoautosomal recessive), hemolytic types of anemia.

# TYPES OF SICKLE CELL ANEMIA

## SICKLE CELL ANEMIA

The homozygous forms of the disease (HgbSS), in which valine, an amino acid, is substituted for glutamic acid at the sixth position of the beta chain.

## SICKLE CELL C DISEASE

A heterozygous variant of sickle cell disease (Hgb SC), characterized by the presence of both HgbS and hemoglobin C (HgbC), in which lysine is substituted for glutamic acid at the sixth position of the beta chains.



- **SICKLE THALASSEMIA DISEASE**

A combination of sickle cell trait and beta thalassemia trait.

Father with sickle cell trait

Mother with sickle cell trait



X



or



or



or



Healthy child

Child with sickle cell trait

Child with sickle cell trait

Child with sickle cell anemia

# INCIDENCE

- Sickle cell anemia is most common in people whose families come from Africa, South or Central America (especially Panama), Caribbean islands, Mediterranean countries (such as Turkey, Greece, and Italy), India, and Saudi Arabia

# PATHOPHYSIOLOGY

- Presence of HbgS
- ↓
- Inflexibility & fragility of the sickle cells
- ↓
- Irreversibly sickle cells cannot pass through the blood vessels
- ↓
- Entanglement & enmeshing of rigid sickle shaped cells with one another blocks the microcirculation, causing vasoocclusion
- ↓
- Increase in the blood viscosity, circulatory stasis & hypoxia leading to further sickling
- ↓
- Absence of blood flow to the tissues
- ↓
- Tissue hypoxia, ischemia, & infarction & extreme pain (vaso-occlusive sickle cell crisis)
- ↓
- Progressive organ failure

# PATHOPHYSIOLOGY

- Sickled RBCs are less able to withstand the stresses of the circulation



- Shortert survival time



- Chronic hemolytic anemia

# CLINICAL FEATURES

## HEART

Chronic hemolytic anemia



Cardiomegally, systolic flow murmur, septal hypertrophy & increased circulation



Impaired contractility



Cardiac decompensation & cardiac failure

# LUNGS

Vasooocclusive crisis



Pulmonary vessels may be occluded by masses of sickled RBCs



Pulmonary insufficiency



Pneumonia



Altered pulmonary function

# SPLLEN

Spleen is enlarged from engorgement with sickled cells



Normal functioning cells are gradually replaced by fibrotic tissues



Spleen is decreased in size & became functional asplenia



Susceptible to infection



# LIVER

Vaso occlusive crisis & rapid destruction of RBCs

↓  
Impairment of hepatic blood flow from anemia & capillary obstruction

↓  
Chronic overproduction of bilirubin

↓  
Chronic hepatic dysfunction

↓  
Jaundice & gall stones

↓  
cirrhosis of liver

# KIDNEYS

Congestion of glomerular capillaries & tubular arterioles with sickle cells & hemosiderin



Tissue hypoxia & necrosis caused by diffuse glomerular & tubular fibrosis



Inability to concentrate urine, hematuria & nephrotic syndrome



Enuresis & polyuria

# CNS

- SCA



- Ischemia & Infarction of the vessels of the CNS caused by vasoocclusion



- Stroke

# EYE

- Peripheral vascular occlusion in the retina
- ↓
- Retinal detachments
- ↓
- Vitreous hemorrhage
- ↓
- Loss of vision

# **SKELETAL SYSTEM**

- Infarction of the bone & bone marrow
- ↓
- Osteoporosis & widening of the medullary Space
- ↓
- Osteoporosis due to weakening of the bone
- ↓
- Skeletal deformities( lordosis & kyphosis)
- ↓
- Osteomyelitis & pathological fracture

# CRISIS

- Vasoocclusive crisis
- Sequestration crisis
- Aplastic crisis
- Hyper hemolytic crisis

# DIAGNOSTIC EVALUATION

- Blood smear-reveal few sickled RBCs
- Sickle turbidity test
- Screening of newborn
- Hemoglobin
- Reticulocyte
- Urine analysis
- Chest X-ray

# MEDICAL MANAGEMENT

## **Hydroxyurea-**

- The first effective drug treatment for adults with severe sickle cell anemia
- Daily doses of the anticancer drug, hydroxyurea, reduced the frequency of painful crises, acute chest syndrome, needed fewer blood transfusions
- Increases production of fetal hemoglobin in the blood



- Electrolyte replacements
- Analgesia
- Antibiotics
- Pneumococcal, H.Influenza & Meningococcal vaccines
- Penicillin prophylaxis

# 1. Minimization of energy expenditure & the use of oxygen

- Bed rest
- PROM
- Assess the child's need for oxygen
- Avoid low oxygen environment
- Decrease physical exertion & emotional stress

## 2.Promote hydration

- Oral fluids & IVF therapy
- Monitor IO
- Evaluate the degree of dehydration
- Observe the s/s of hypokalemia
- Monitor serum electrolytes
- Avoid overheating
- Encourage to take more fluids

# 3.Replacement of electrolytes & blood

Replacement of electrolytes to correct metabolic acidosis

Blood transfusion

Exchange transfusion

Observe the s/s of transfusion reaction

Observe the complications of multiple blood transfusion

Chelation therapy

# 4. Relief of pain

- Analgesics
- Heat applications
- Do not give cold compress

# 5. Administration of antibiotics

- Antibiotics
- Pneumococcal, H.Influenza & Meningococcal vaccines
- Penicillin prophylaxis

# 6. Management by surgical interventions

- Splenectomy
- Monitor abdominal girth
- Monitor v/s & BP
- Cholecystectomy

## **7.PSYCHOSOCIAL ISSUES**

**Require regular medical attention** -Especially before and after operations, dental extraction and during pregnancy.

**Adherence to medical regimen**-Vitamins, antibiotics, fluid intake, activity |Schools must be involved

**Family planning**

**Suitable types of employment**

**Air travel**-Increased fluids, pain killers or oxygen may be recommended

### **LIFE STYLE MODIFICATION**

Healthy diet.

Folic acid

Should drink at least 8 glasses of water every day

Regular physical activity -avoid exercise that makes you very tired

Sleep and rest

Avoid extremes of heat and cold

Be cautious at high altitudes



# 8. Observation for complication

- Infections
- Shock
- Profound anemia
- CHF
- Splenic rupture
- Gall stones
- Stroke
- Bone changes

# PREVENTION

- Public education
- Screening-antenatal screening  
(amniocentesis, & fetoscopy with fetal  
blood sampling)
- Genetic counseling
- Birth control measures.

1. Correcting the “defective gene” and inserting it into the bone marrow
2. Turning off the defective gene and simultaneously reactivating another gene that turns on production of fetal hemoglobin.

No real cure for Sickle Cell Anemia at this time.

“In the past 30 years, the life expectancy of people with sickle cell anemia has increased. Many patients with sickle cell anemia now live into their mid-forties and beyond.”

# CLUB FOOT













Normal



Clubfoot  
in baby





# DEFINITION

- A clubfoot is a complex deformity of a foot & ankle that includes forefoot adduction, midfoot supination, hindfoot varus and ankle equinas.
- Congenital clubfoot involves bone deformity & malposition with soft tissue contracture.

- A **clubfoot**, or congenital talipes equinovarus (CTEV),-is a congenital deformity involving one foot or both

# INCIDENCE

- Clubfoot occurs in about 1 in 700 to 1 in 1000 births.
- In half of the cases (50%), both feet are usually affected.
- Over 220,000 children, in the developing world are born each year with clubfoot.
- This occurs in males more often than in females by a ratio of 2:1

# TERMINOLOGY

- **Talipes-** Foot & ankle
- **Varus-** Bending inward
- **Valgus-** Bending outward
- **Equinus-** Toes are lower than the heel
- **Calcaneus-** Toes are higher than the heel

# TYPES OF CLUBFOOT

## Talipes varus

- Heels being turned inward from the midline of the leg

## **Talipes valgus**

- Heels being turned outward from the midline of the leg



## **Talipes equinovagum**

- Heels being elevated & turned outward from the midline of the body

## **Talipes calcaneovarus**

- Heels being turned toward the midline of the body and the anterior part of the foot being elevated.

# CAUSES

- Idiopathic
- Arrested fetal development
- Abnormal neuromuscular dysfunction
- Intrauterine compression from oligohydrominos or from amniotic band Syndrome
- myelomeningocele (spina bifida) or arthrogryposis
- Breech presentation

# SIGNS & SYMPTOMS

- walks on the outside of his foot which is not meant for weight-bearing.
- skin breaks down
- chronic ulceration
- infection.

# DIAGNOSTIC EVALUATION

- Prenatal ultrasonography
- Hip examination
- Anteroposterior and lateral radiography
- Observation

# MANAGEMENT

## **Involves 3 stages**

- Correction of the deformity
- Maintenance of the correction until normal muscle balance is recognised
- Follow up observation to avert possible recurrence of the deformity

- Clubfoot is treated with manipulation by podiatrists, physiotherapists, orthopedic surgeons, specialist Ponseti nurses, or orthotists by providing braces to hold the feet in orthodox positions, serial casting, or splints called knee ankle foot orthoses (KAFO).

## Casting

- A series of plaster or fibreglass casts are applied to the foot and lower limb - these are replaced every few weeks, which each cast progressively moving the foot towards a more corrected position.
- The number of times the cast needs to be replaced will be determined by the severity of the clubfoot



## **Strapping and physiotherapy –**

- Strips of adhesive strapping are passed around the foot, up the sides of the leg, and over the top of the knee, to hold the foot in a corrected position.
- This is usually done weekly, following some physiotherapy.

## **Plaster fixation**

- The surgeon manipulates the foot into position, and holds it in place with plaster.
- The plasters tend to stay on for about 4 weeks, or changed weekly

## **Ponseti Method**

- It involves regular casting, followed by the wearing of a splint.
- splint may be needed at night for 2 to 4 years, which makes this a long term treatment,
- Follow up care needed

# **SURGICAL MANAGEMENT**

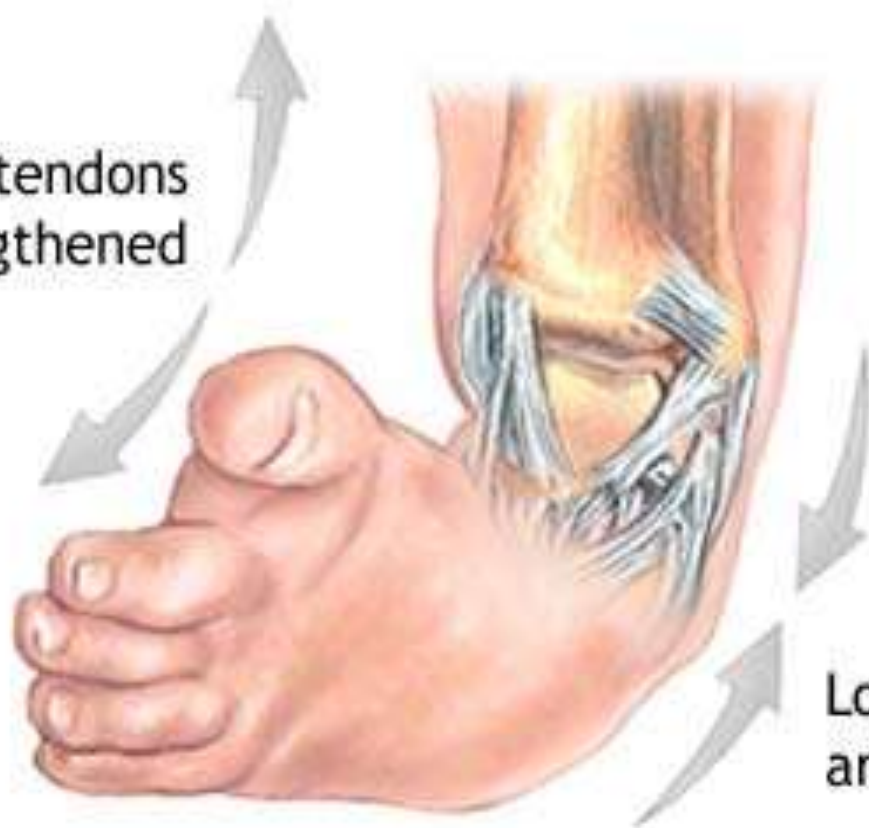
variety of surgical procedures which may be done in isolation or in combination:

- Tenotomy
- Anterior Tibial Tendon Transfer

- Dennis-Brown bars with straight last boots,i.e, ankle foot orthoses



Short tendons  
are lengthened



Long tendons  
are shortened



Before



After

Before



After





# NURSING MANAGEMENT

- Maintain position
- ice packs
- Circulation & skin around the cast and the toes should be checked frequently
- care of the cast, corrective shoes and splint
- Physical therapy
- Divertional therapy
- Close follow-up –regular foot examination

# complication

- Skin irritation & ulceration
- Neuromuscular impairment
- Difficulty in walking

# OUTCOME

- The outcome depends on the type and degree of deformity, the age when treatment was begun, & the success of the treatment.

# FRACTURE

- A **child bone fracture** is a medical condition in which a bone of a child is cracked or broken



# CAUSATIVE FACTORS

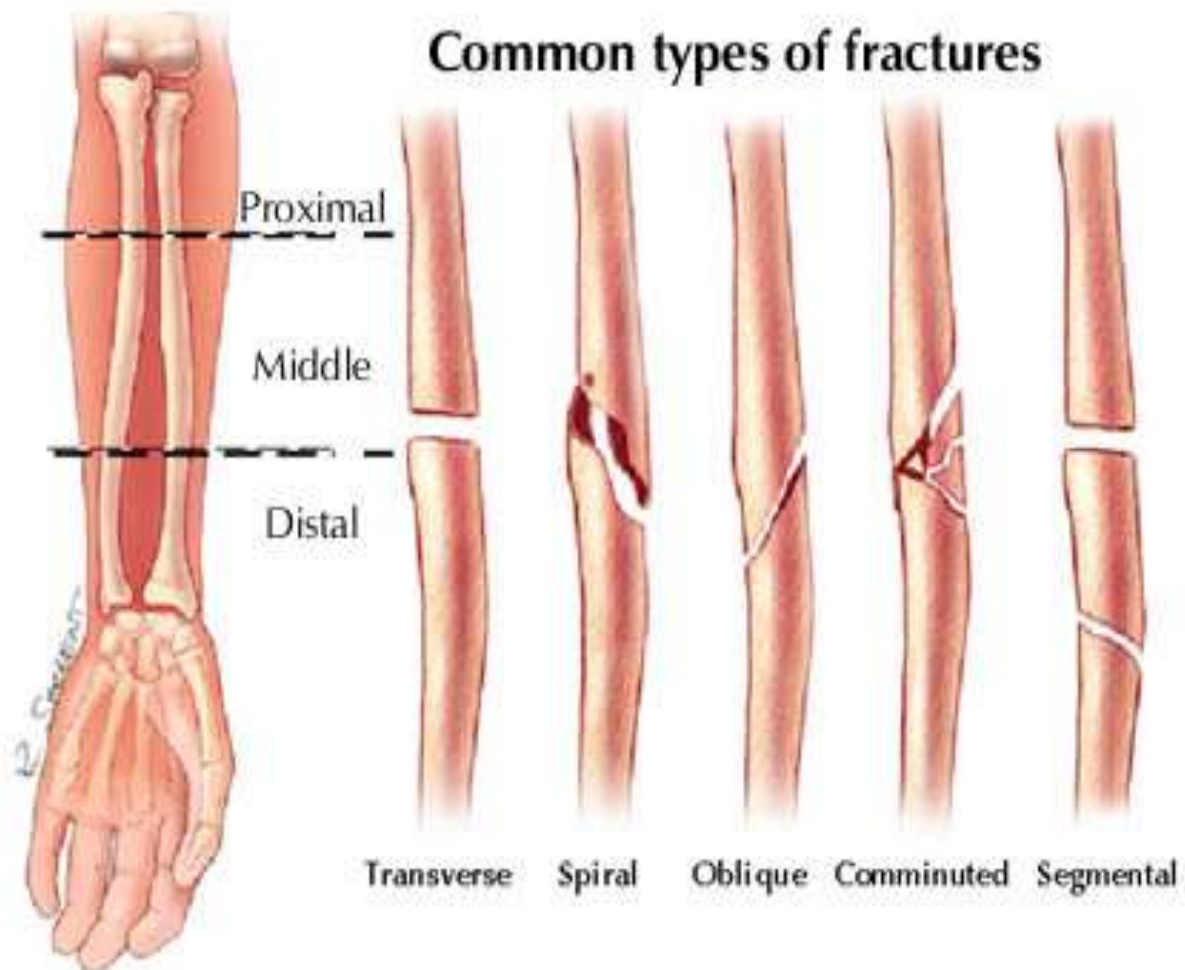
- Trauma at birth
- Accidents by falling, climbing
- Child abuse
- Risk taking behavior
- Rough handling, twisting & pulling the

# FEATURES OF FRACTURE IN CHILDREN

- Growth plates
- Thick periosteum
- Plastic bone
- Rapid healing
- Stiffness is not usual
- Response to injury

# CLASSIFICATION

- Complete fracture





- Bends –at 45 degree
- Greenstick fracture

# CLINICAL FEATURES

- Swelling
- Pain
- Tenderness
- Diminished functional use of affected extremities

## **5 Ps**

- Pain & point of tenderness
- Pulselessness
- Pallor
- Parasthesia
- Paralysis

# DIAGNOSTIC EVALUATION

- History & physical examination
- X-ray
- Hb & Hct
- WBC
- Myoglobin MB

# MANAGEMENT

## GOAL

- To reestablish alignment & length of the bony fragments
- To retain alignment & length
- To restore function to the injured parts
- To prevent further injury

# METHODS OF FRACTURE REDUCTION

- Closed reduction
- Traction
- Open reduction

# **NURSING MANAGEMENT**

## **CAST**

- Prevention of circulatory, neurologic & respiratory disturbances
- Maintenance of body temperature
- Maintenance of skin integrity & Prevention of infection
- Maintenance of cast integrity & cleanliness

- Promotion of appropriate muscle activity
- Provision of comfort measures
- Prevention of urinary stasis & constipation
- Provision of support during cast removal
- Education of the parents & their child



# **TRACTION**

- Reduce muscle spasms
- Align dislocated bones
- Prevent or treat contracture deformities

# NURSING DIAGNOSIS

complication

# Developmental Dysplasia of the Hip

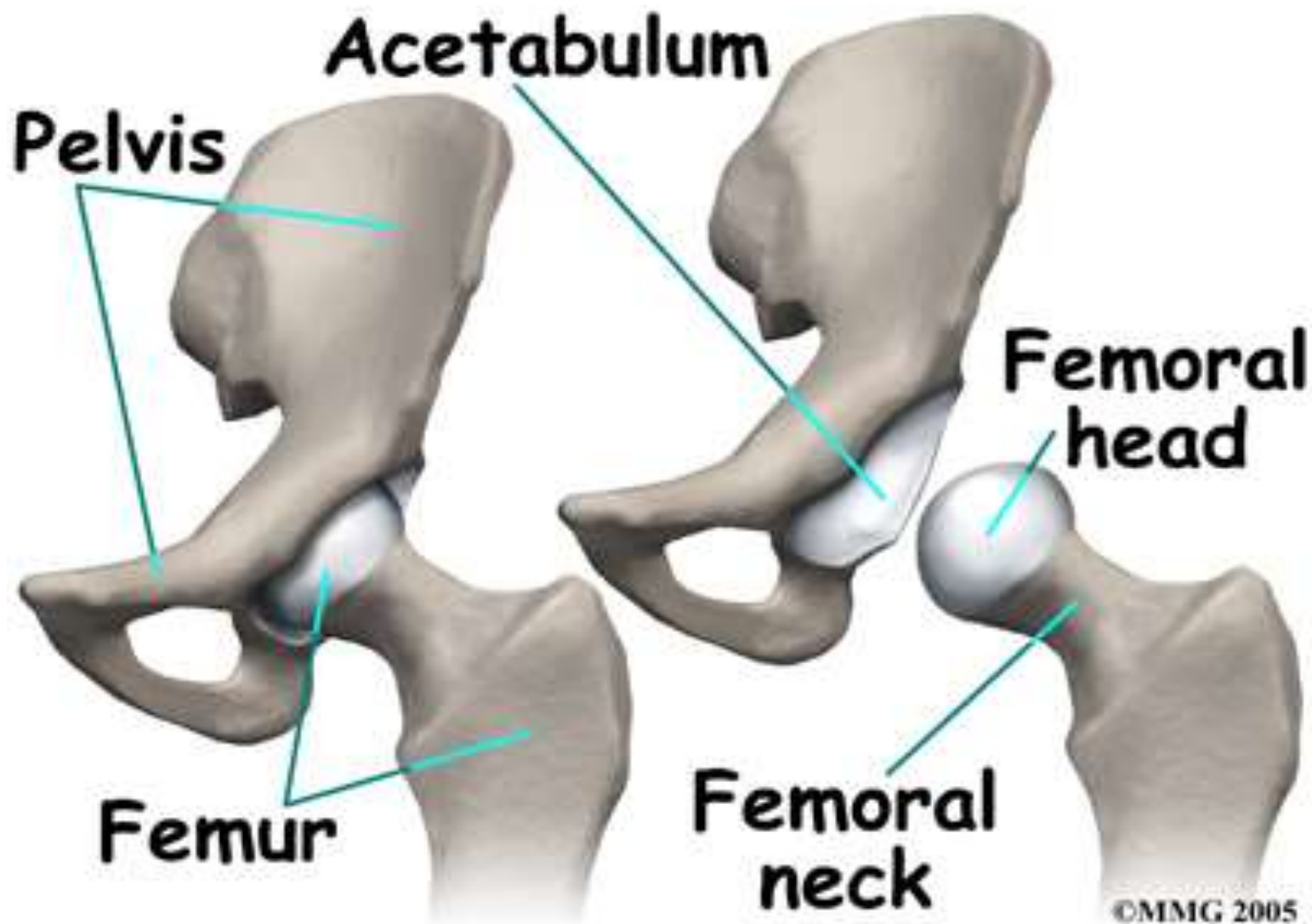


**Normal  
hip joint**



**Dysplastic  
hip joint**







**Femoral  
head**

**Femoral  
neck**

**Greater  
trochanter**

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# DEFINITION

- DDH / CDH is a condition involving the abnormal development of the proximal femur and /or acetabulum.
- DDH describes a spectrum of disorders related to abnormal development of the hip that may develop at anytime during fetal life, infancy & childhood.



# INCIDENCE

- 1 in 750 live births
- Left hip is most commonly affected (60%)  
right hip(20%); both (20%)
- Girls are affected 8 times more than boys
- 30-50% of infants with DDH are born breech
- Caucasian children have higher incidence

# ETIOLOGY

- Hereditary
- Female baby
- Intrauterine positioning
- Breech positioning
- Neurological disorders
- Meningomyelocele

- Cerebral palsy
- Multiple fetus
- Oligohydrominos Large fetal size
- intrauterine malpositioning or crowding

# **PATHOPHYSIOLOGY**

## **A) ACCORDING TO STRUCTURE INVOLVED**

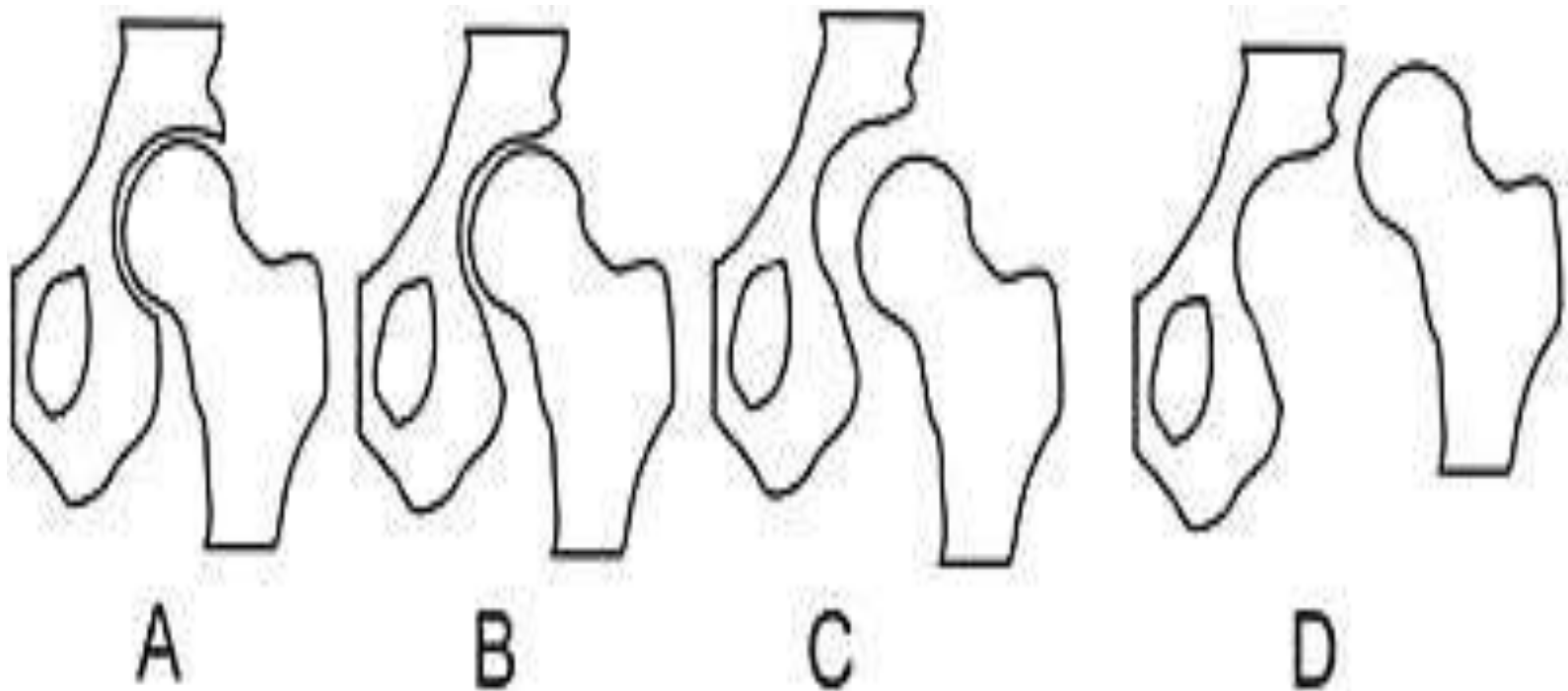
- i) Typical-neurological intact
- li) Terotological-involves neuromuscular defect such as arthrogyposis & myelodysplasia

- **B) ACCORDING TO DEGREE OF DDH**
- i) **Dysplasia**-delay in acetabular development
- ii) **Subluxation**- head of the femur is partially displaced from its normal position in the acetabulum
- iii) **Diallocation**-Head of the femur loses contact with the acetabulum

# Types of misalignments of femur head to socket in hip

A: Normal. B: Dysplasia. C: Subluxation.

D: Luxation



# CLINICAL MANIFESTATIONS

## INFANTS

### Allis / Galeazzi sign

- One knee lower than the other when the infant is on the back with the knees flexed.





# Galeazzi Test

Difference  
in  
knee  
height



**Barlow  
test**

**Ortolani  
test**



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- Asymmetry of the thigh
- Presence of gluteal & knee folds
- Diminished spontaneous movements
- Shortening of the affected legs & externally rotated
- Inability to abduct the knee fully
- Posterior pulging of femoral head

## **OLDER INFANT AND CHILD**

- Affected leg is shorter
- Trendelenburgs sign
- Lordosis
- Waddling gait
- Walking is delayed with the presence of pain in the joints

# DIAGNOSTIC EVALUATION

- Initial examination at birth
- Ortoloni or barlow test
- Shortened limb on the affected side
- Assymetry gluteal and thigh folds
- Broadening of the perineum

- MRI
- Arthrograms
- Radiographic examination
- CT scan

# MANAGEMENT

- **EARLY INFANCY**
- Splinting
- Pavlik harness
- Plaster hip spica
- Double or triple diaper
- Protective abduction brace

- CAST



# Cast care instructions

- Keep the cast clean and dry.
- Check for cracks or breaks in the cast.
- Rough edges can be padded to protect the skin from scratches.
- Do not scratch the skin under the cast by inserting objects inside the cast.
- Never blow warm or hot air into the cast.



- Do not put powders or lotion inside the cast.
- Cover the cast during feedings to prevent spills from entering the cast.
- Prevent small toys or objects from being put inside the cast.
- Elevate the cast above the level of the heart to decrease swelling.
- Do not use the abduction bar on the cast to lift or carry the baby.

# PAVLIK HARNESS



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- *Pavlik harness* can be used for three to nine months.
- It keeps the hip in flexion and abduction.
- It may be worn until the doctor can no longer move the hip in and out of the socket.
- In the older child, x-rays may be used to confirm that the hip is stable.

# TRACTION

- . Traction consists of pulleys, strings, weights, and a metal frame attached over or on the bed.
- The purpose of traction is to stretch the soft tissues around the hip and to allow the femoral head to move back into the hip socket.
- Traction is most often used for approximately 10 to 14 days

## **Later infancy/toddler period**

- Traction
- Closed reduction and open reduction of the hip
- Spica cast

## **Older children**

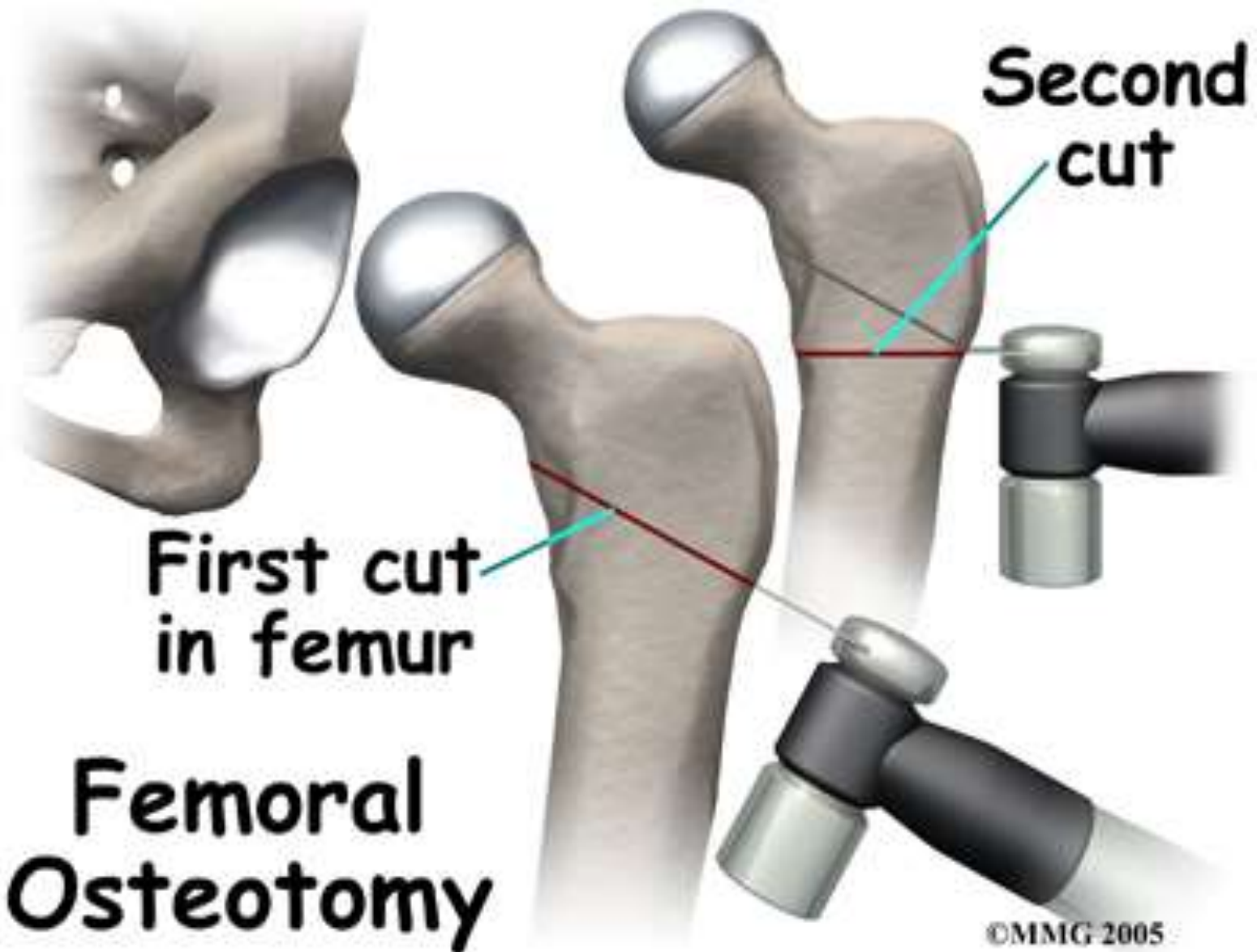
- Preoperative traction
- Tonotomy
- Osteotomy



# Pelvic Osteotomies







# Femoral Osteotomy



**Femoral head  
realigned**

**Femoral  
head  
reattached**



# NURSES RESPONSIBILITY

- Maintaining the correct position of the hip
- Caring of a child in a cast
- Providing optimal nutrition
- Encourage normal physical, emotional & social development
- Educating the parents
- Observing complications

# COMPLICATIONS

- Avascular necrosis
- Loss of ROM of the affected hip
- Leg length discrepancy
- Early osteoarthritis
- Femoral nerve palsy
- Recurrent dislocation
- Unstable hip

# PROGNOSIS

- Prognosis is good if early diagnosis is made and treatment with good care is given

# NURSING DIAGNOSIS

**THANK YOU**