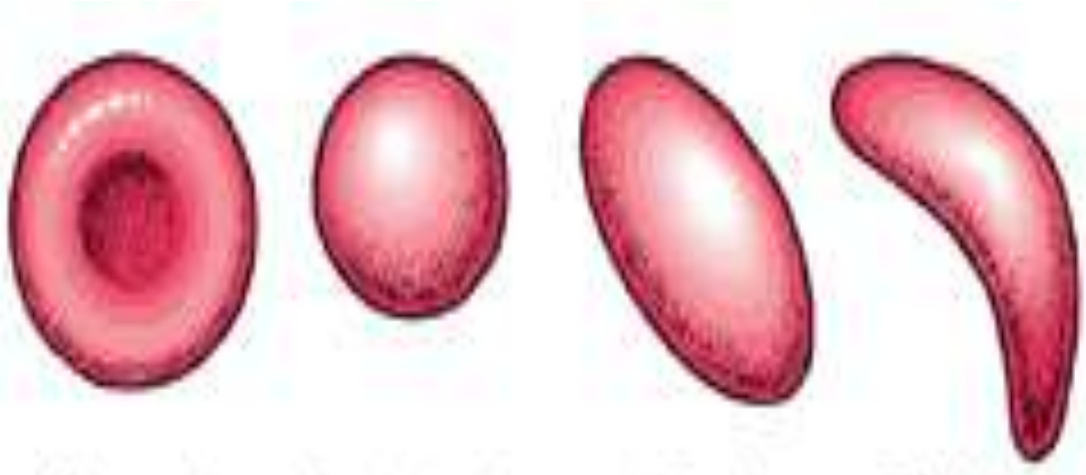
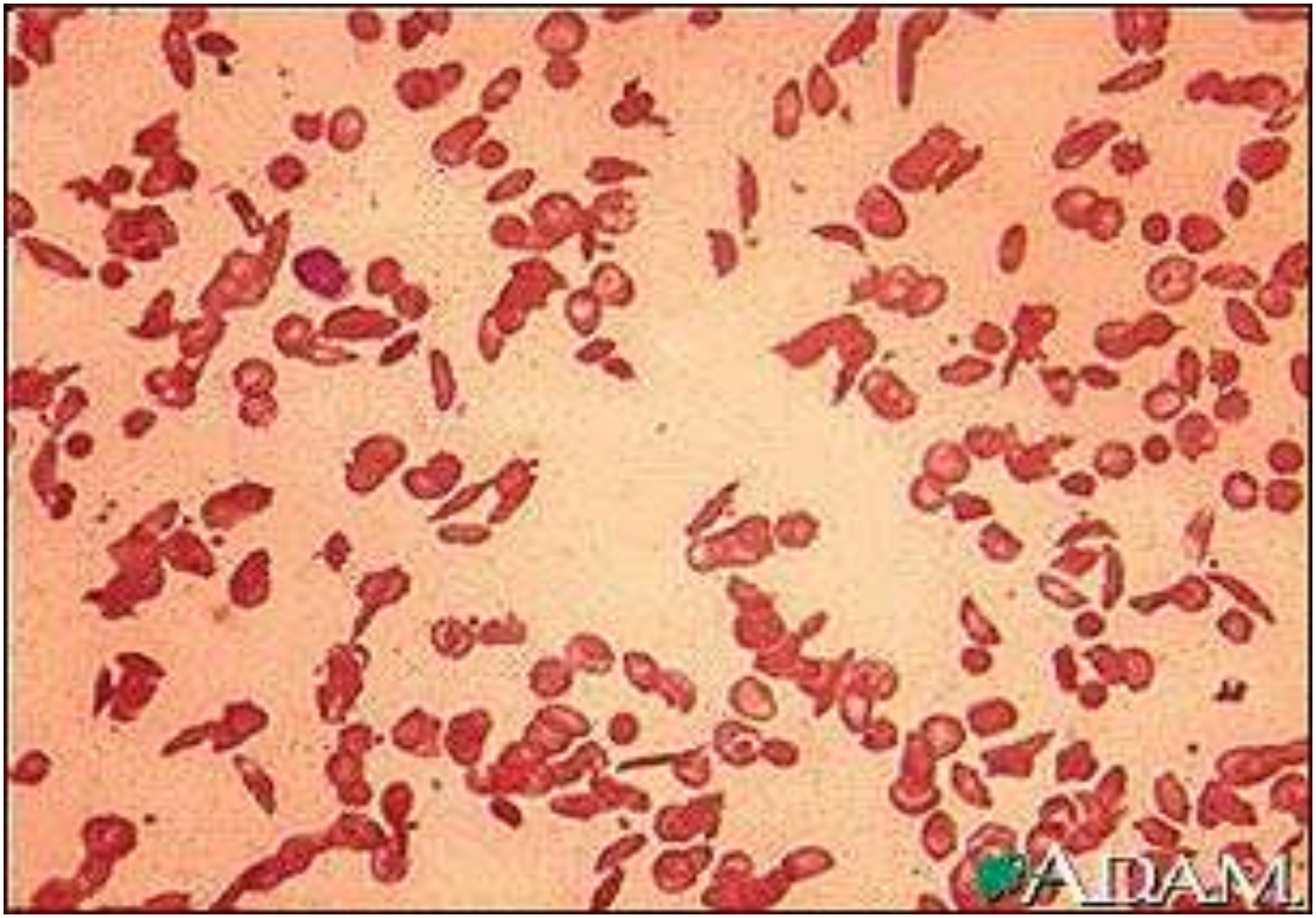
SICKLE CELL ANEMIA

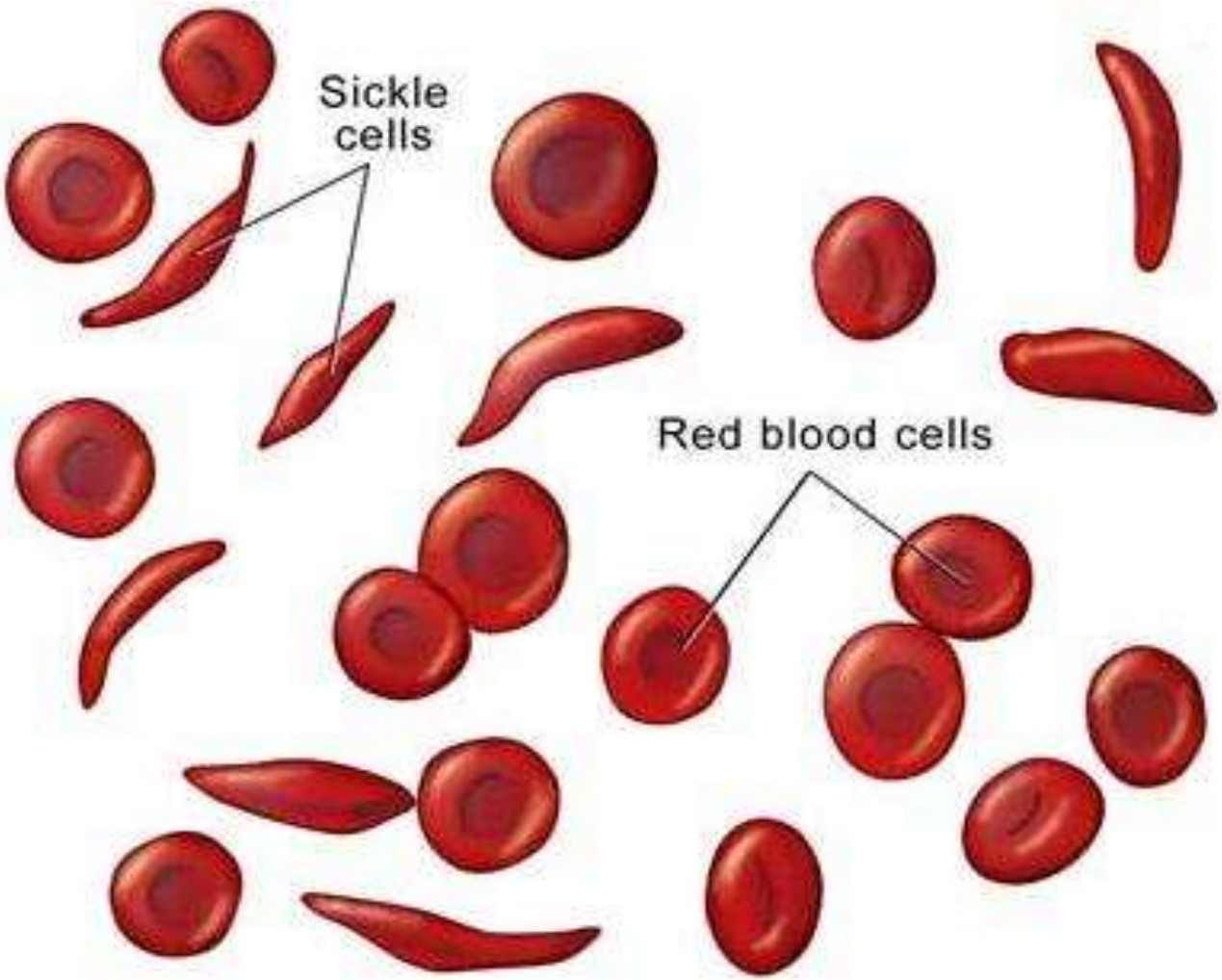

Normal cell
Spherical cell
Oval cell
Sickle cell

Normal red blood cell

Sickled red blood cell


A.D.A.M.

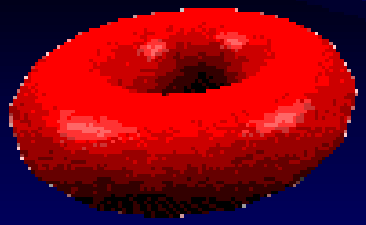

A.D.A.M.


Sickle cells
Red blood cells

Normal vs. Sickle Hemoglobin

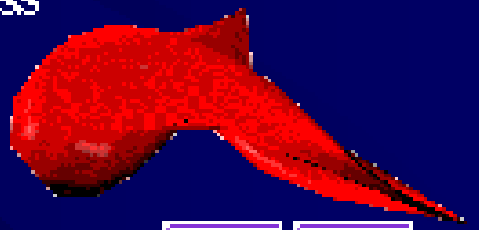
Normal

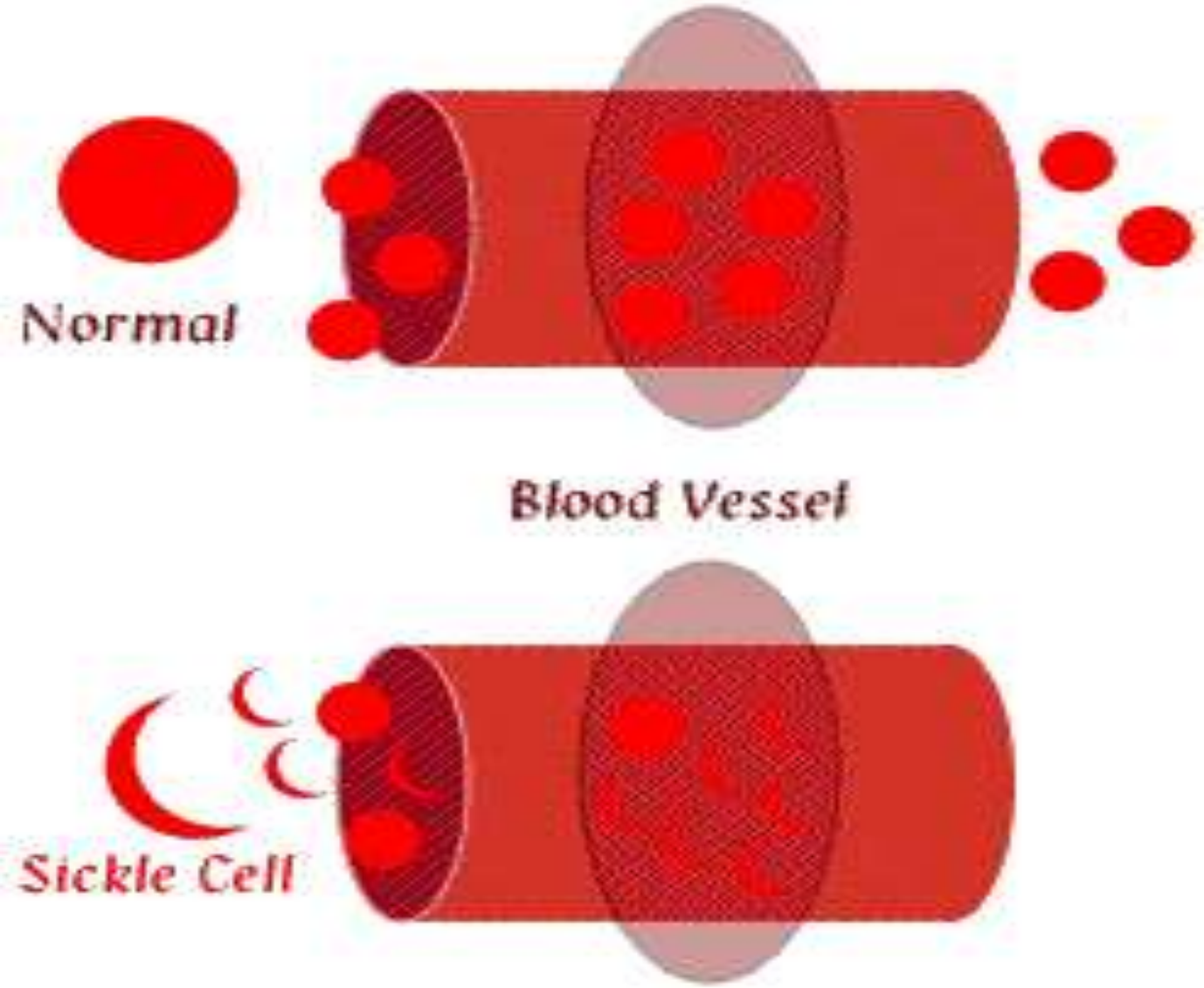
- disc-Shaped
- soft(like a bag of jelly)
- easily flow through small blood vessels
- lives for 120 days



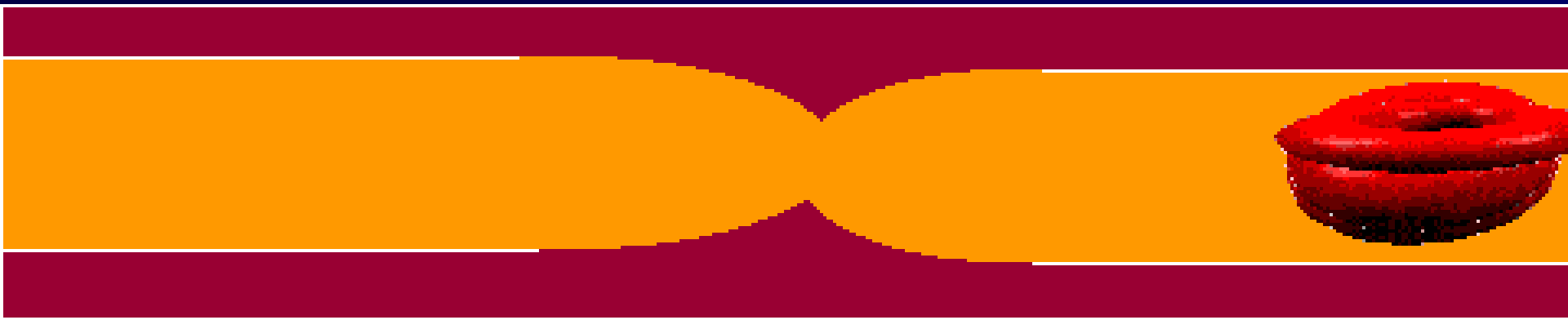
Sickle

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

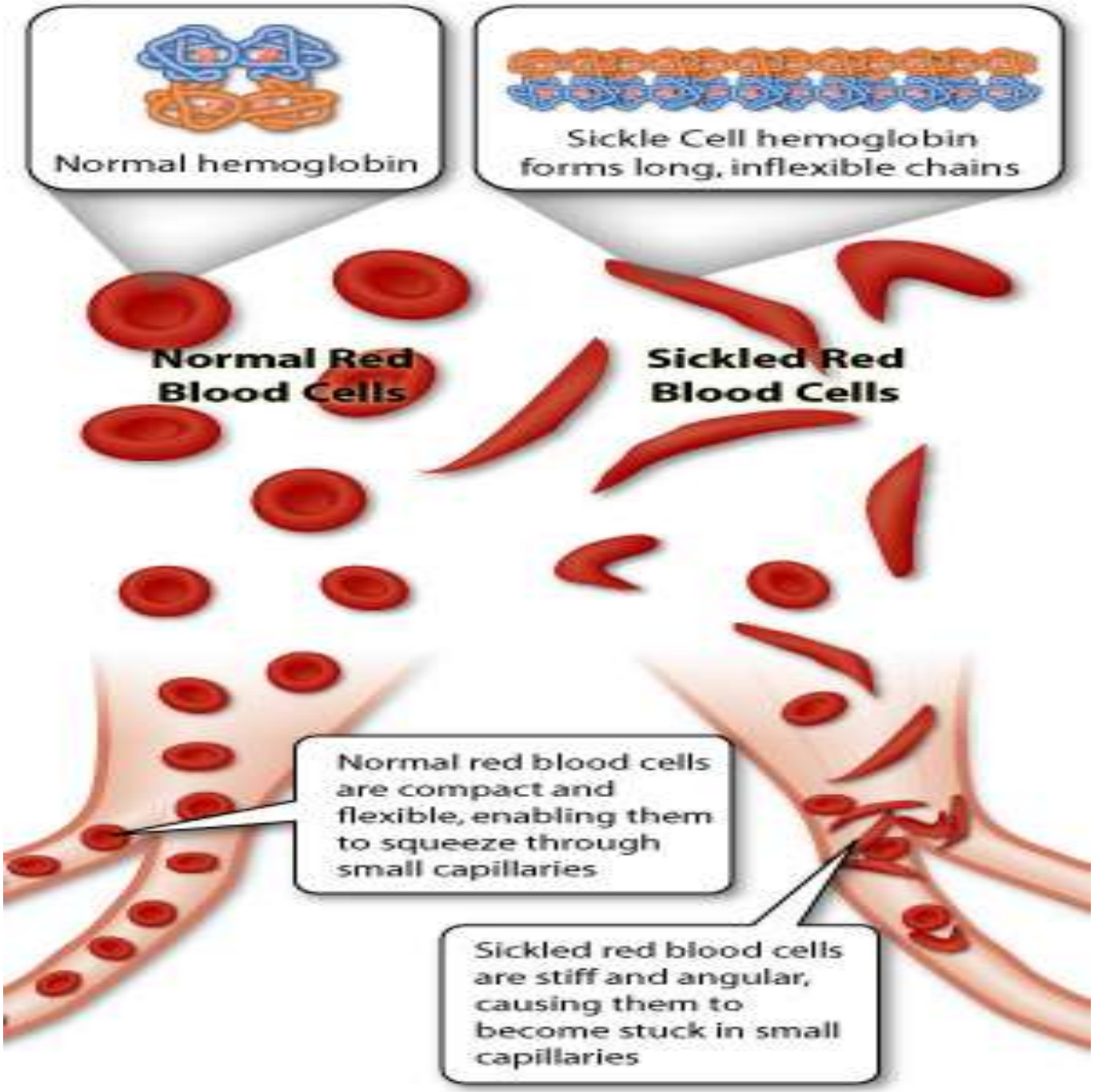


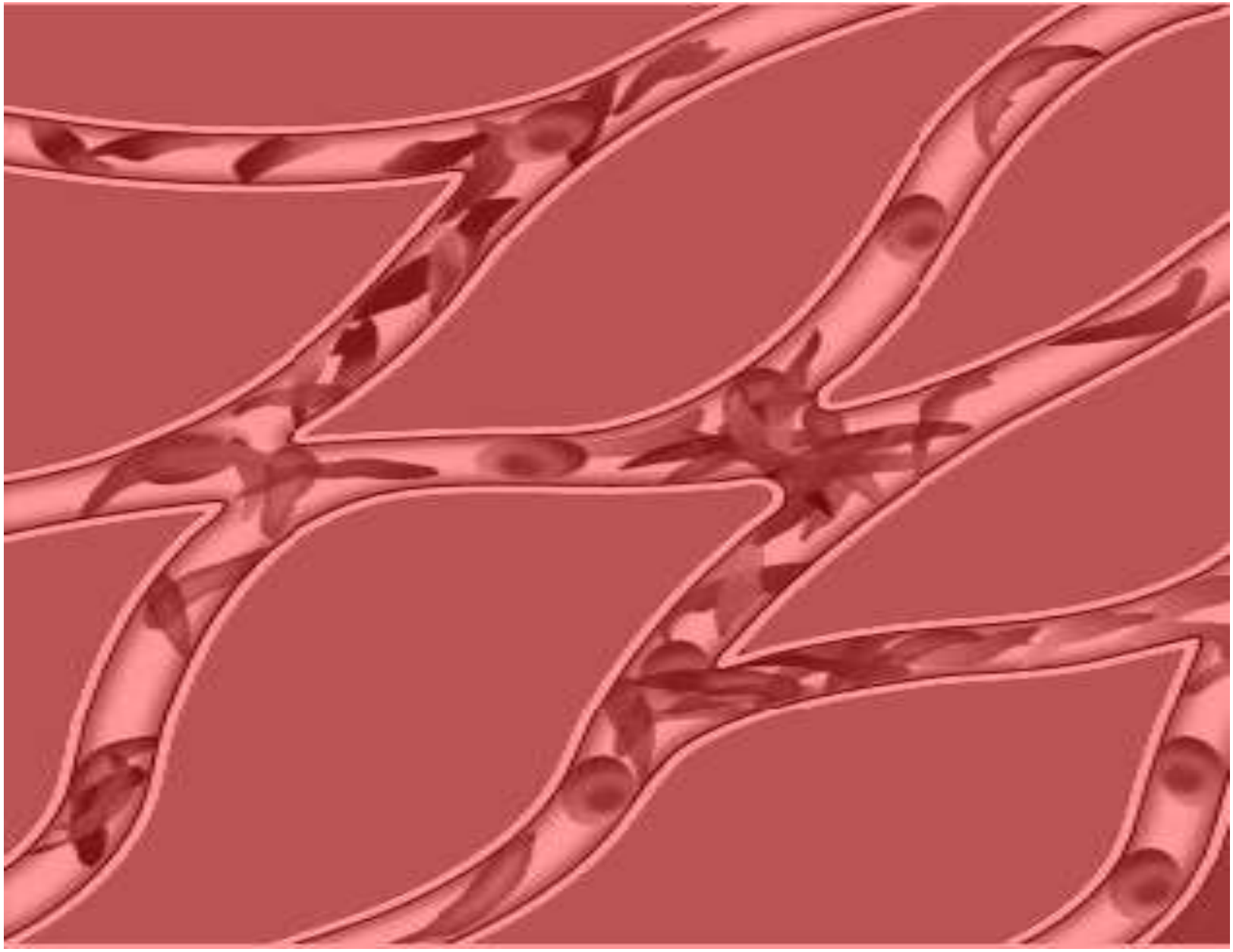

Normal
Blood Vessel
Sickle Cell

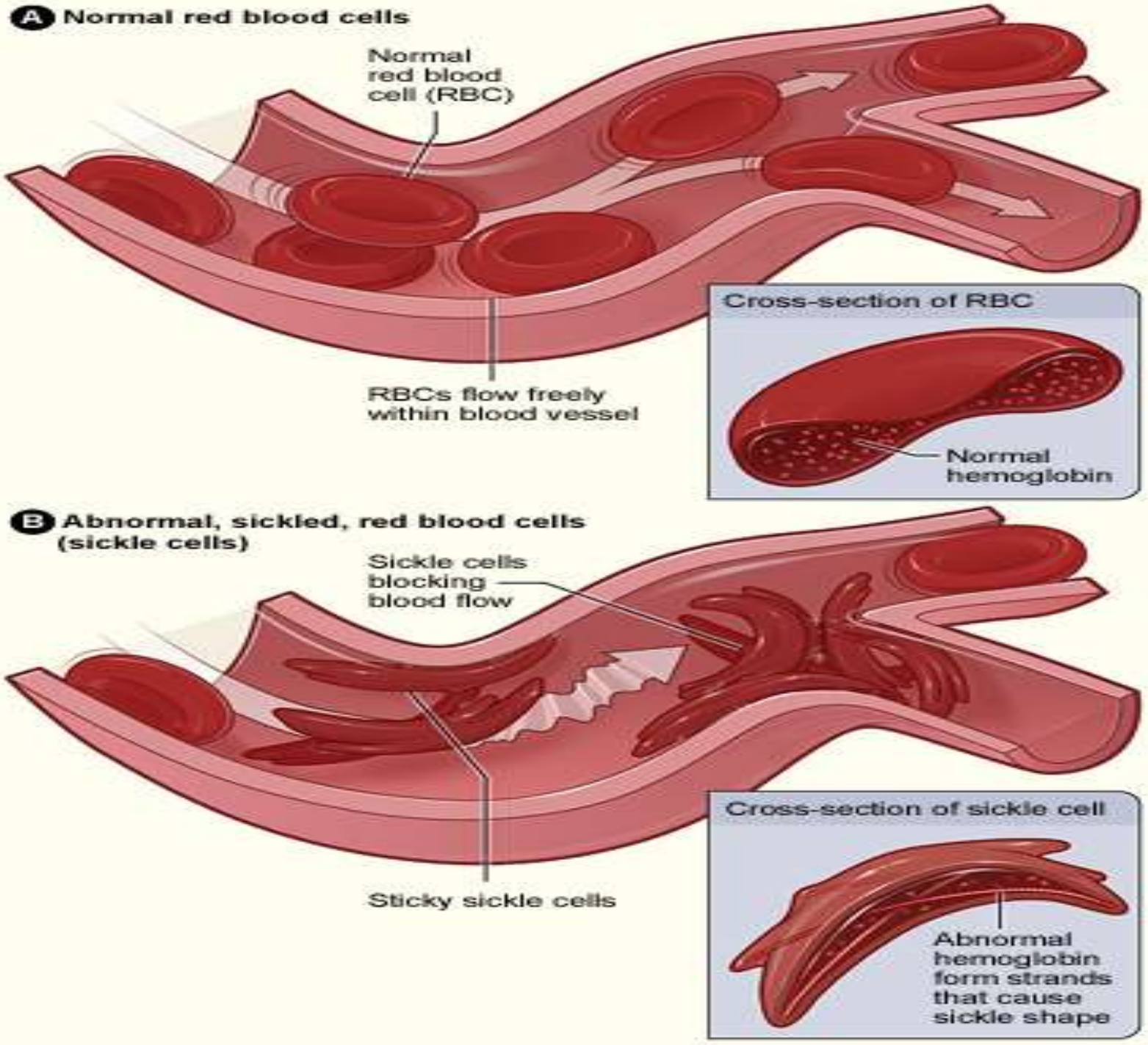
Normal vs. Sickle red cells



If no oxygen, then pain and damage occurs

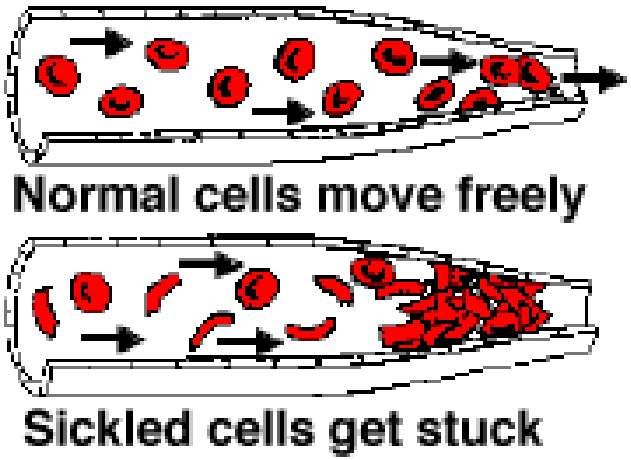

Normal hemoglobin
Sickle Cell hemoglobin forms long, inflexible chains
Normal Red Blood Cells
Sickled Red Blood Cells
Normal red blood cells are compact and flexible, enabling them to squeeze through small capillaries
Sickled red blood cells are stiff and angular, causing them to become stuck in small capillaries




A Normal red blood cells
Normal red blood cell (RBC)
RBCs flow freely within blood vessel
Cross-section of RBC
Normal hemoglobin
B Abnormal, sickled, red blood cells (sickle cells)
Sickle cells blocking blood flow
Sticky sickle cells
Cross-section of sickle cell
Abnormal hemoglobin form strands that cause sickle shape

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 hemoglibinopathies are hereditary, genetically determined (autoautosomal recessive), hemolytic types of anemia.

TYPES OF SICKLE CELL ANEMIA

SICKLE CELL ANEMIA

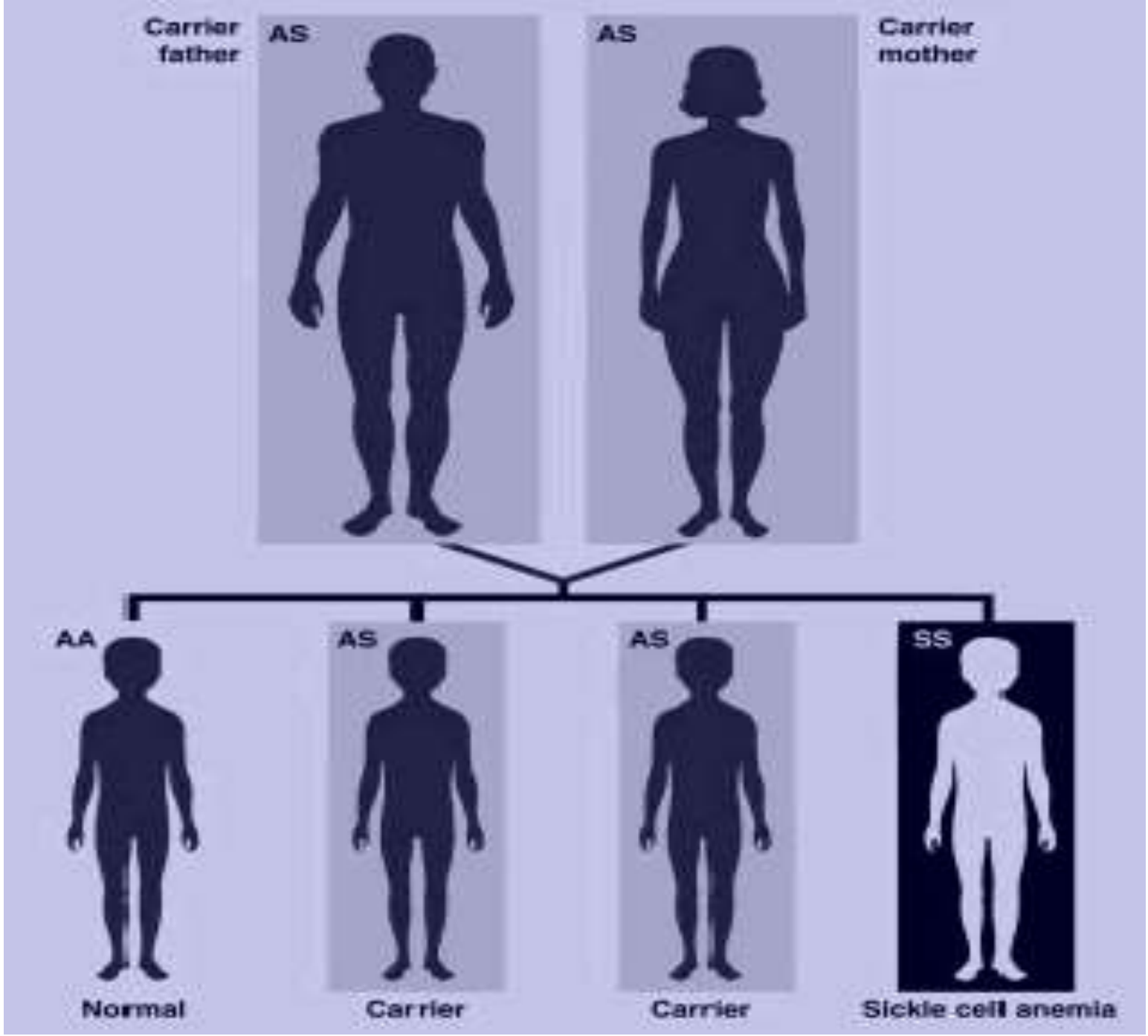
The homozygous forms of the disease(HgbSS), in which valine, an aminoacid, is substituted for glutamic acid at the sixth position of the beta chain.

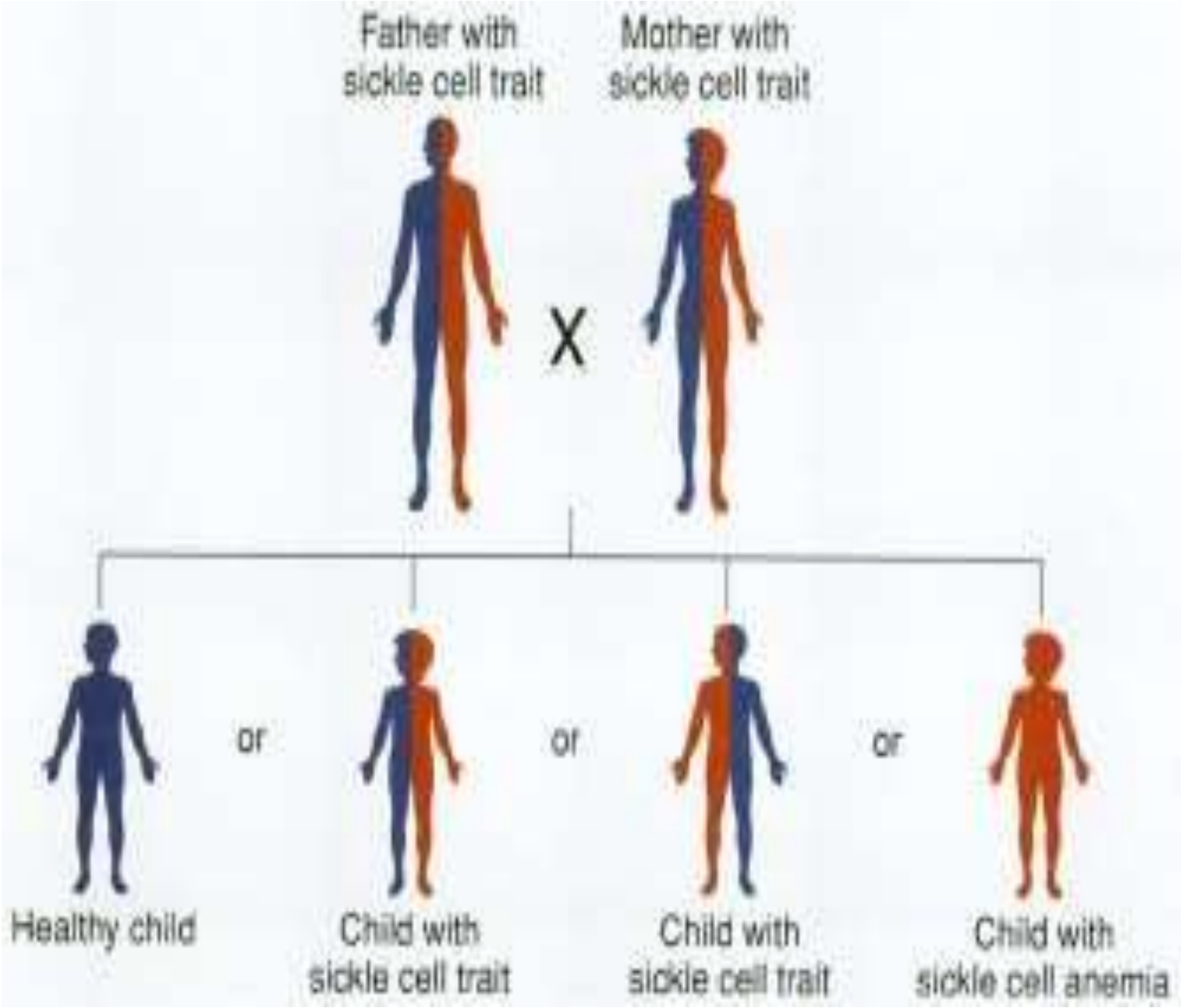
SICKLE CELL C DISEASE

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.


Carrier father
AS
AS
Carrier mother
AA
AS
AS
SS
Normal
Carrier
Carrier
Sickle cell anemia


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

PATHOPHYSIOLOGY

- Presence of HbgS
- Inflexibility & fragility of the sickle cells
- Irrevesibily sickle cells cannot pass through the blood vesels
- Entanglement & enmeshing of rigid sickle shaped cells with one another blocks the microcirculation,causing vasoocclusion
- Increase in the blood viscosity, circulatory stasis & hypoxia lending to further sickling
- Absence of blood flow to the tissues
- Tissue hypoxia,ischemia,& infarction & extreme pain(vaso-occlusive sicle 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

Vasoocclusive 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
- Osteiporesis & widening of the medullary Space
- Osteoporosis due to weakening of the bone
- Skeletal deformities(lordosis & kyposis)
- 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 childs 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.PSYCHOSOCAIAL ISSUES

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

Adherence to medical regimen-Vitamins, antibiotics, fluid intake, activity lSchools must be involved

Family planning

Suitable types of employment

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

8.Observation for complication

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

PREVENTION

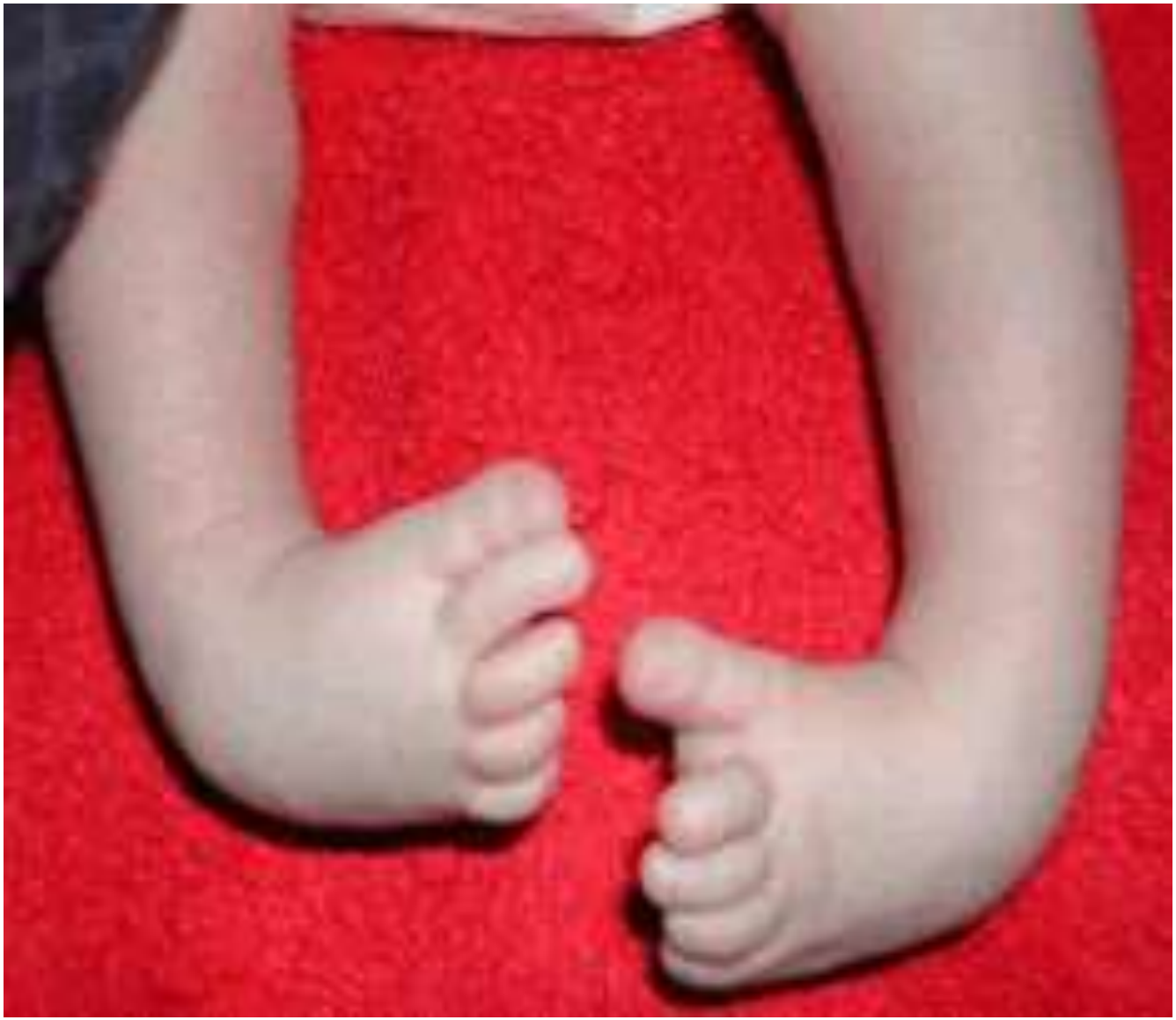
- Public education
- Screening-antenatal screening (amniocentesis,& fetoscopy with fetal blood sammpling)
- Genetic counseling
- Birth control measures.

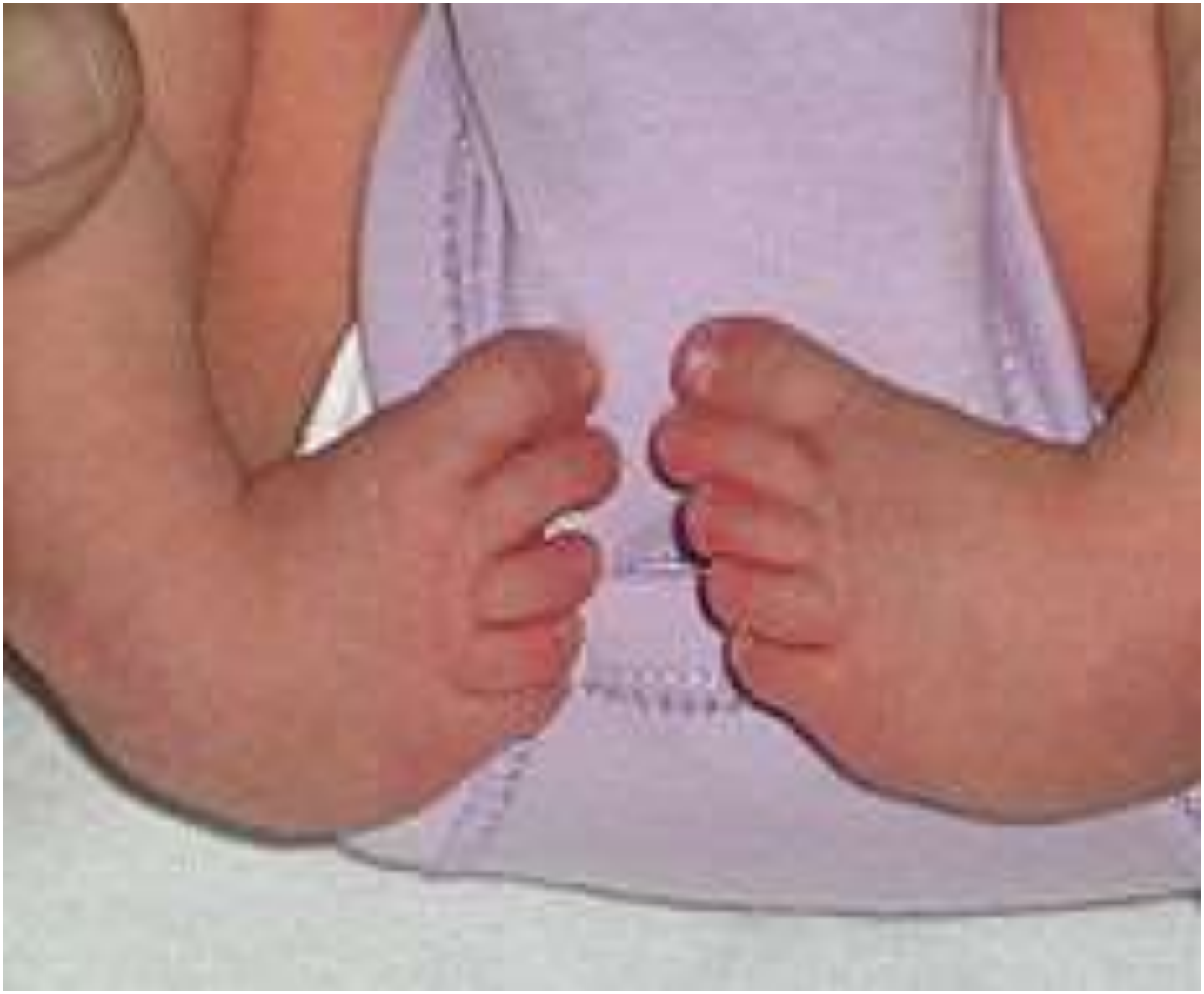
1. Correcting the “defective gene” and inserting it into the bone marrow
2. Turning off the defective gene and simultaneously reactivating another gene that turns on production of fetal hemoglobin.

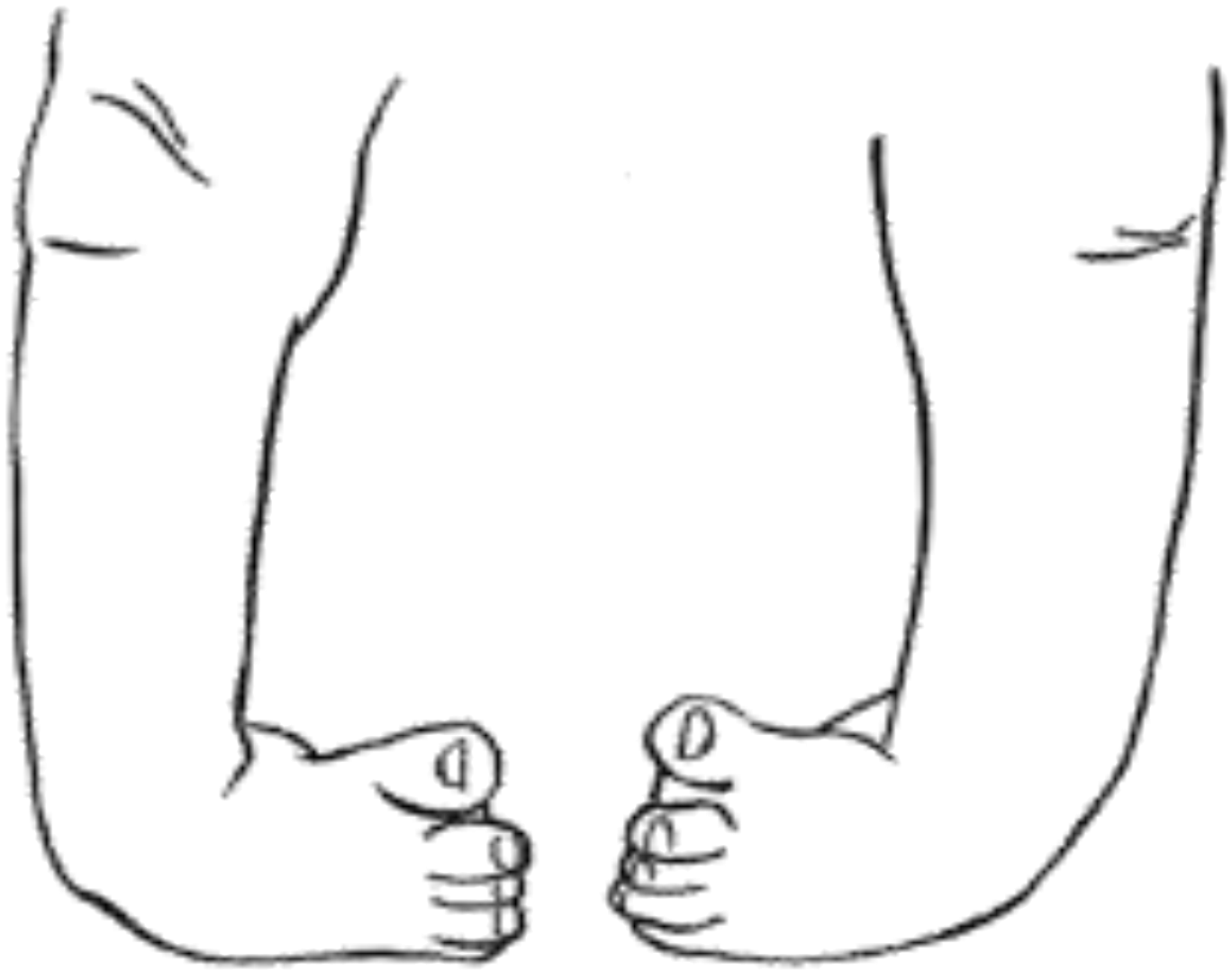
No real cure for Sickle Cell Anemia at this time.

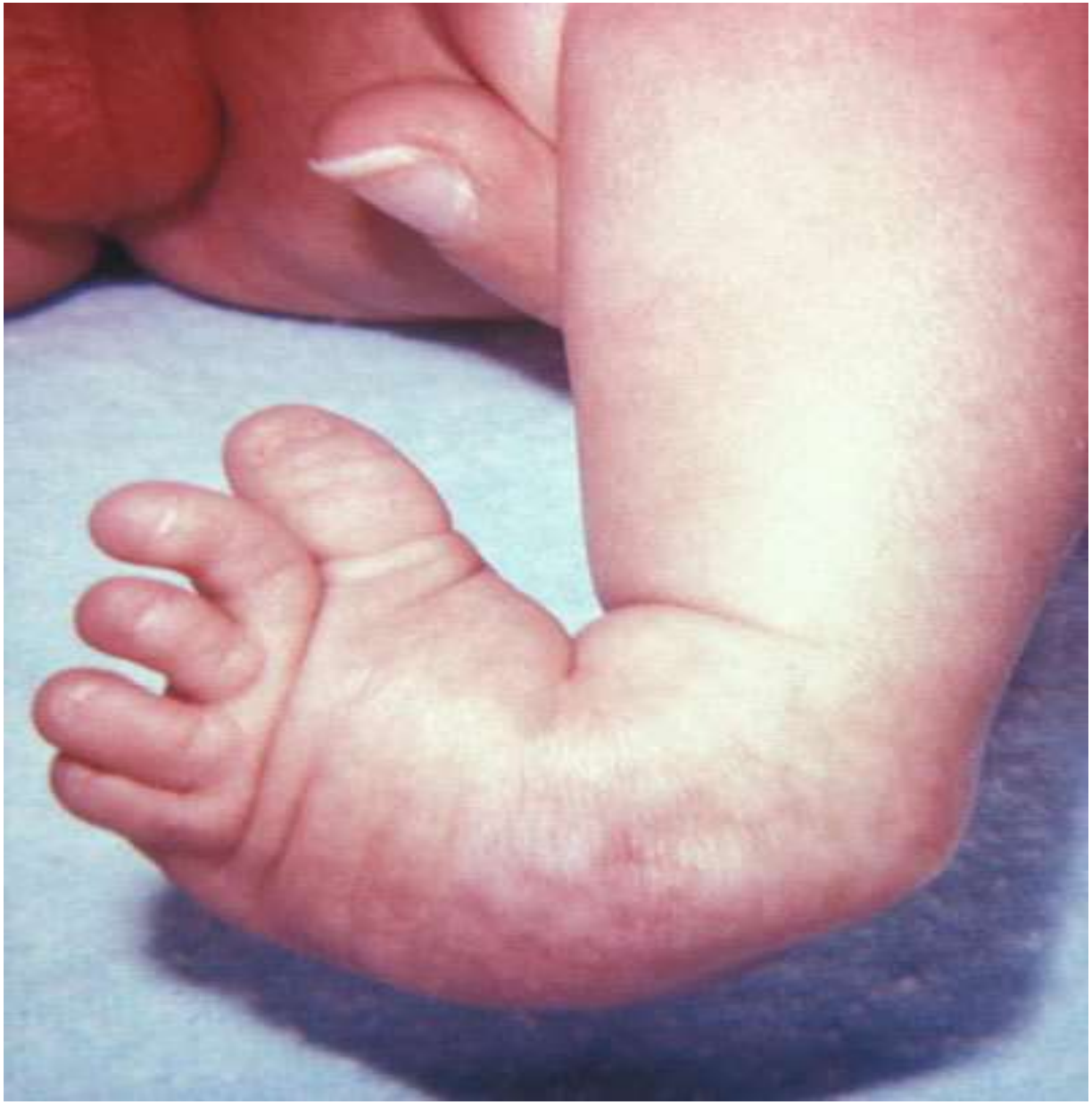
“In the past 30 years, the life expectancy of people with sickle cell anemia has increased. Many patients with sickle cell anemia now live into their mid-forties and beyond.”

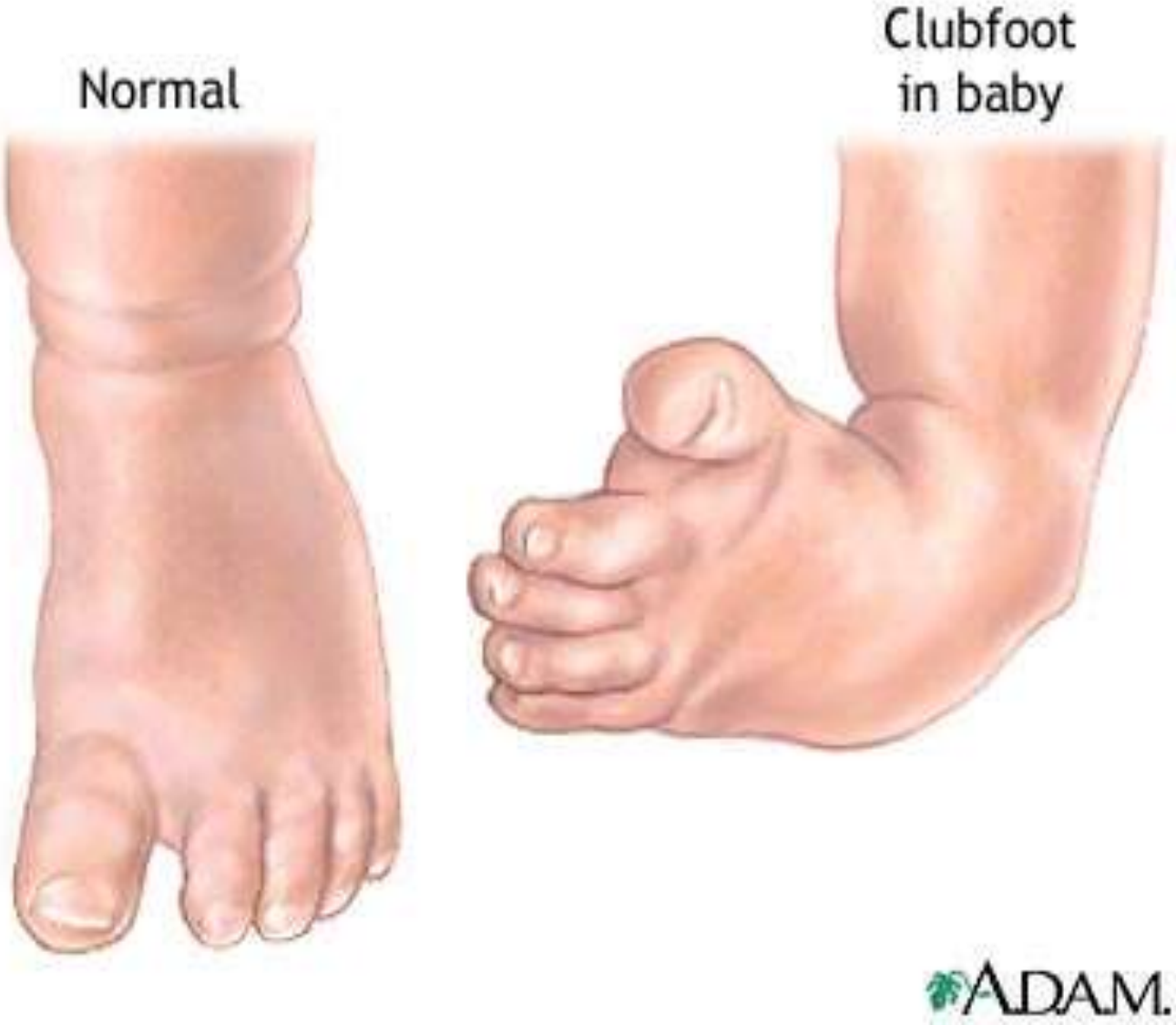
CLUB FOOT










Normal
Clubfoot
in baby
A.D.A.M.

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 in1000 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

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

TYPES OF CLUBFOOT

Talipus varus

- Heels being turned inward from the midline of the leg

Talipus valgus

- Heels being turned outward from the midline of the leg

Talipus equinovalgus

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

Talipus 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
- Anterioposterior 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

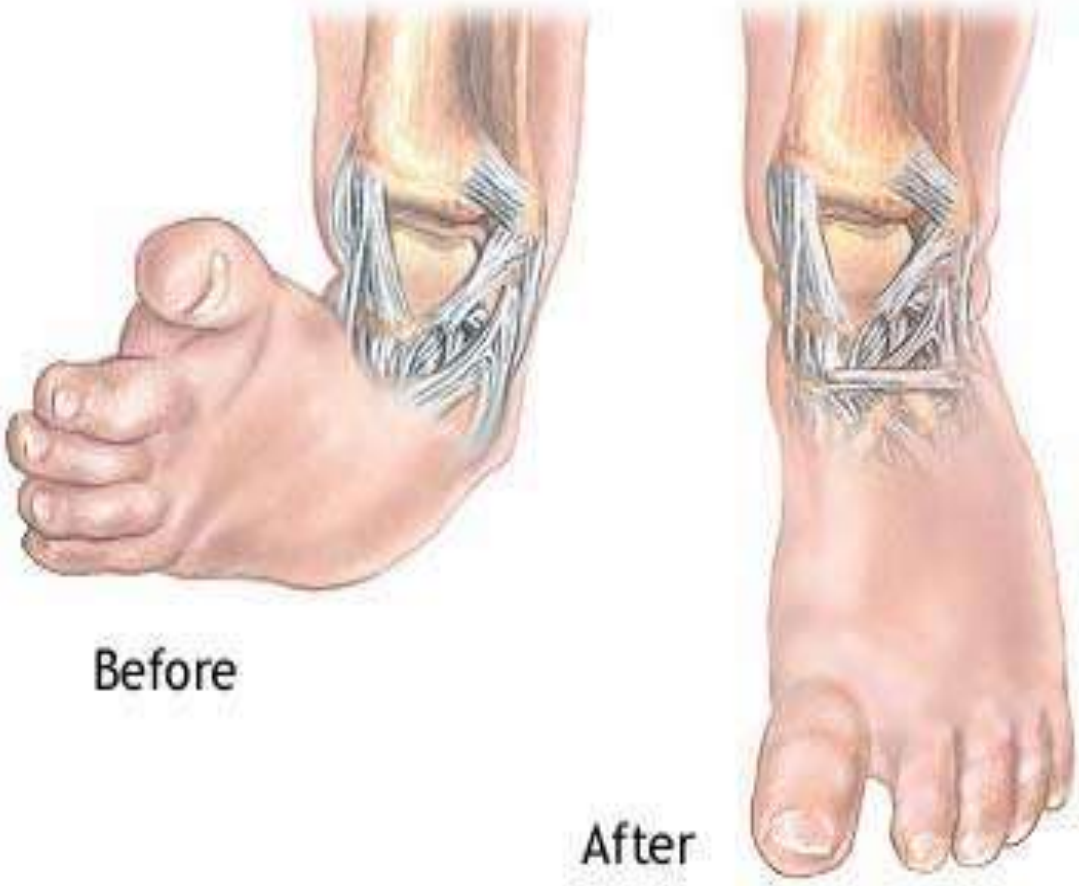
SURGICAL MANAGEMENT

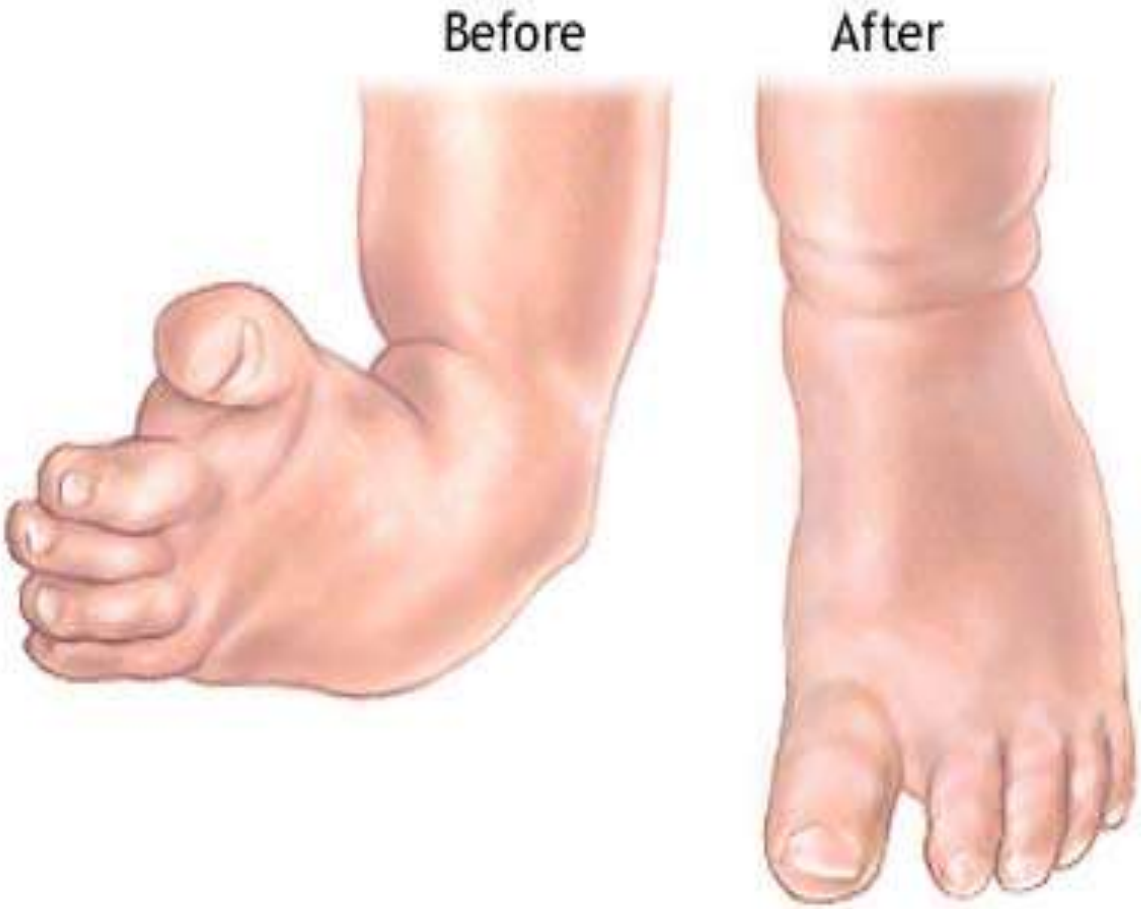
variety of surgical procedures which may be done in isolation or in combination:

- 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
A.D.A.M.





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 impairement
- 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

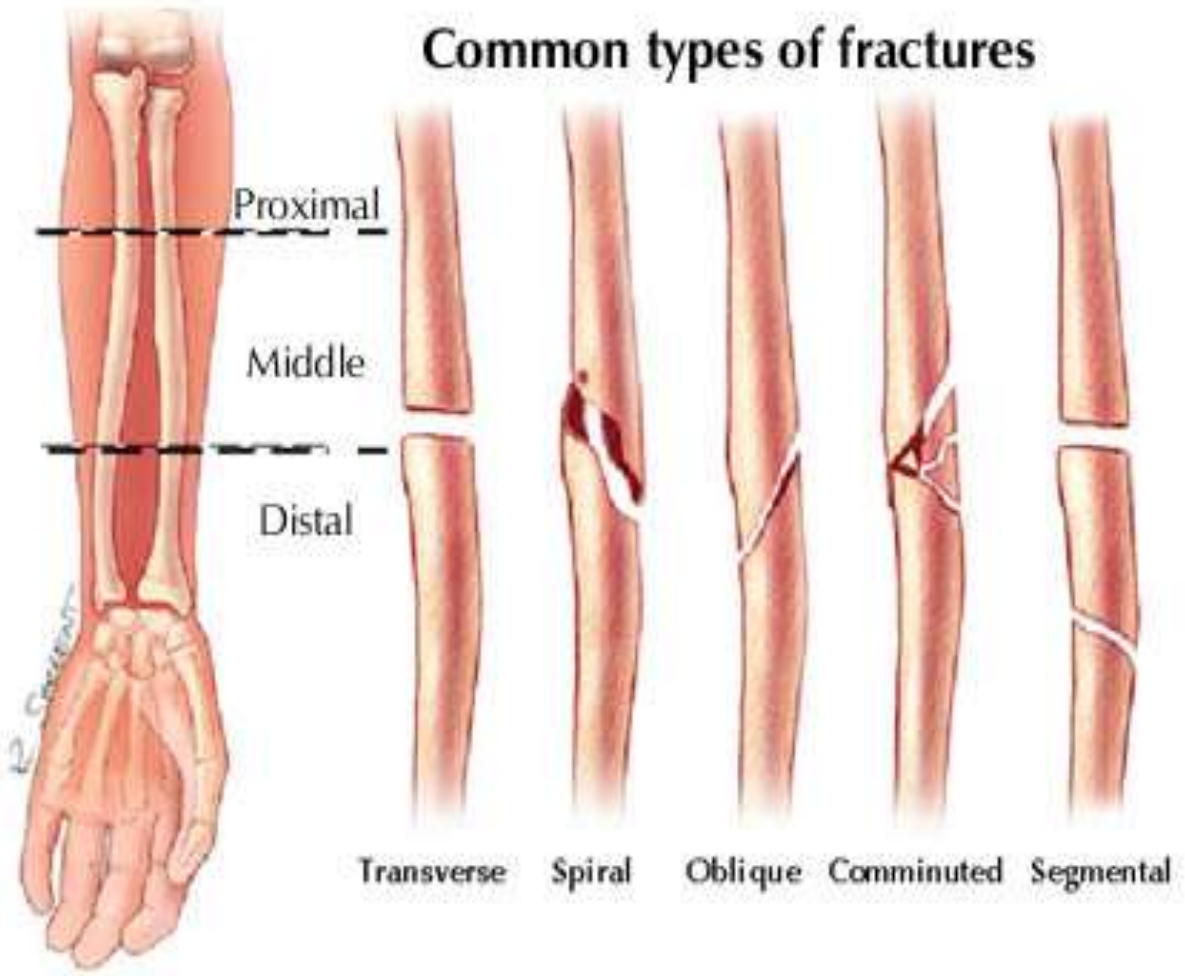
- Trauma at birth
- Accidents by falling, climbing
- Child abuse
- Risk taking behavior
- Rough handling, twisting & pulling the

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 disturbancces
- Maintanace of body temperature
- Maintanance of skin integrity & Prevention of infection
- Maintanace of cast integrity & clenliness

- 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

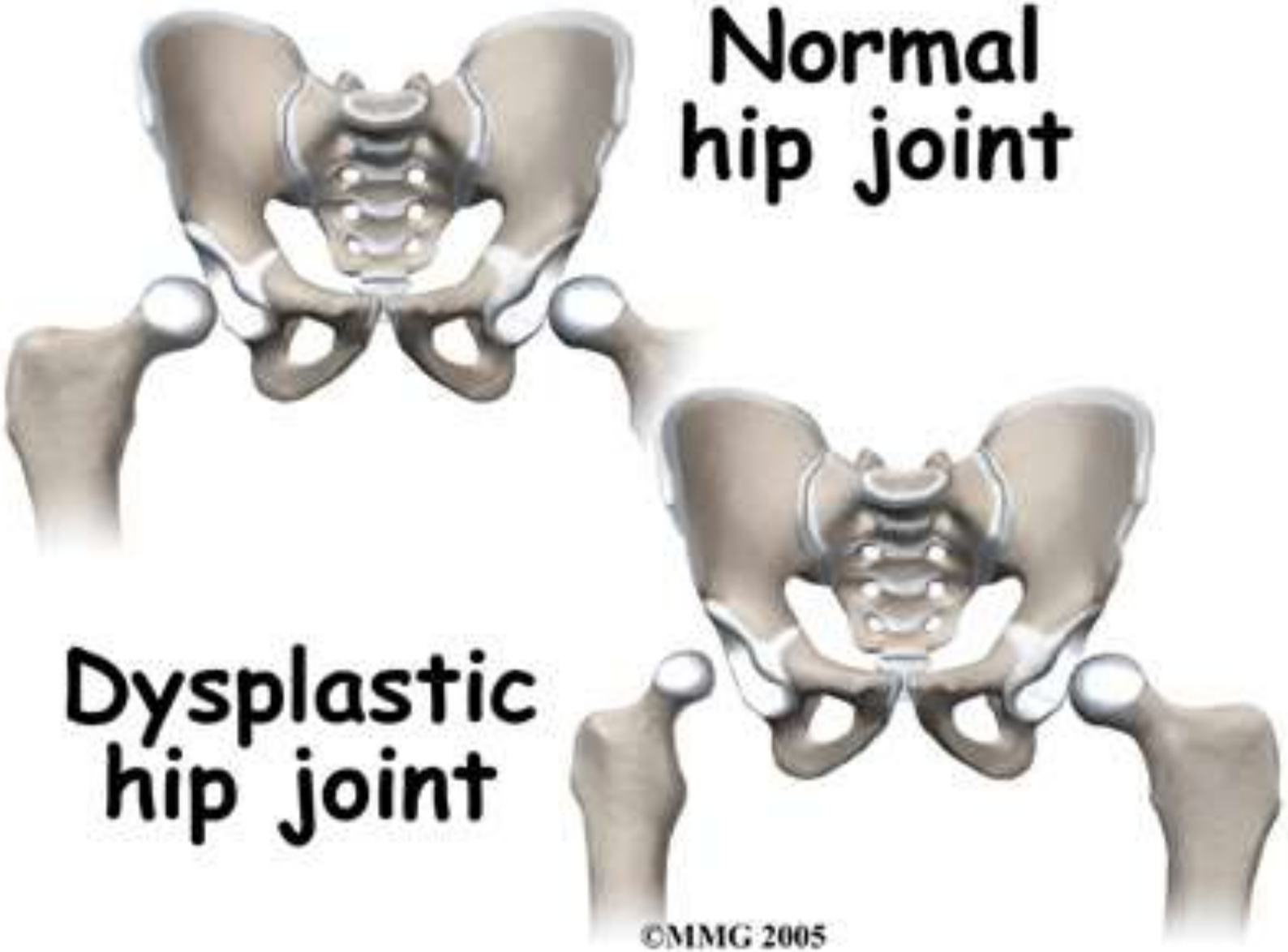
TRACTION

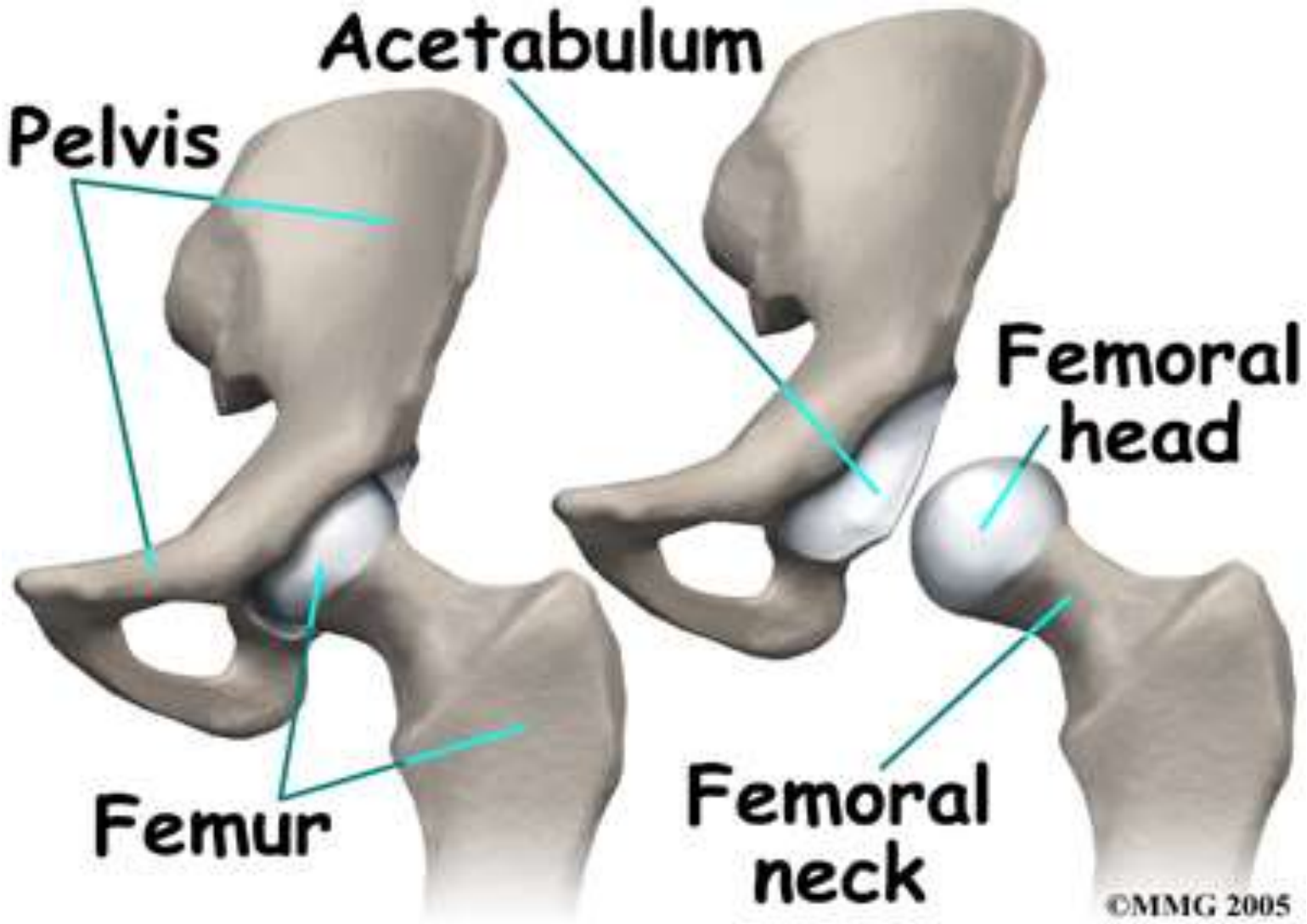
- Reduce muscle spasms
- Align dislocated bones
- Prevent or treat contracture deformities

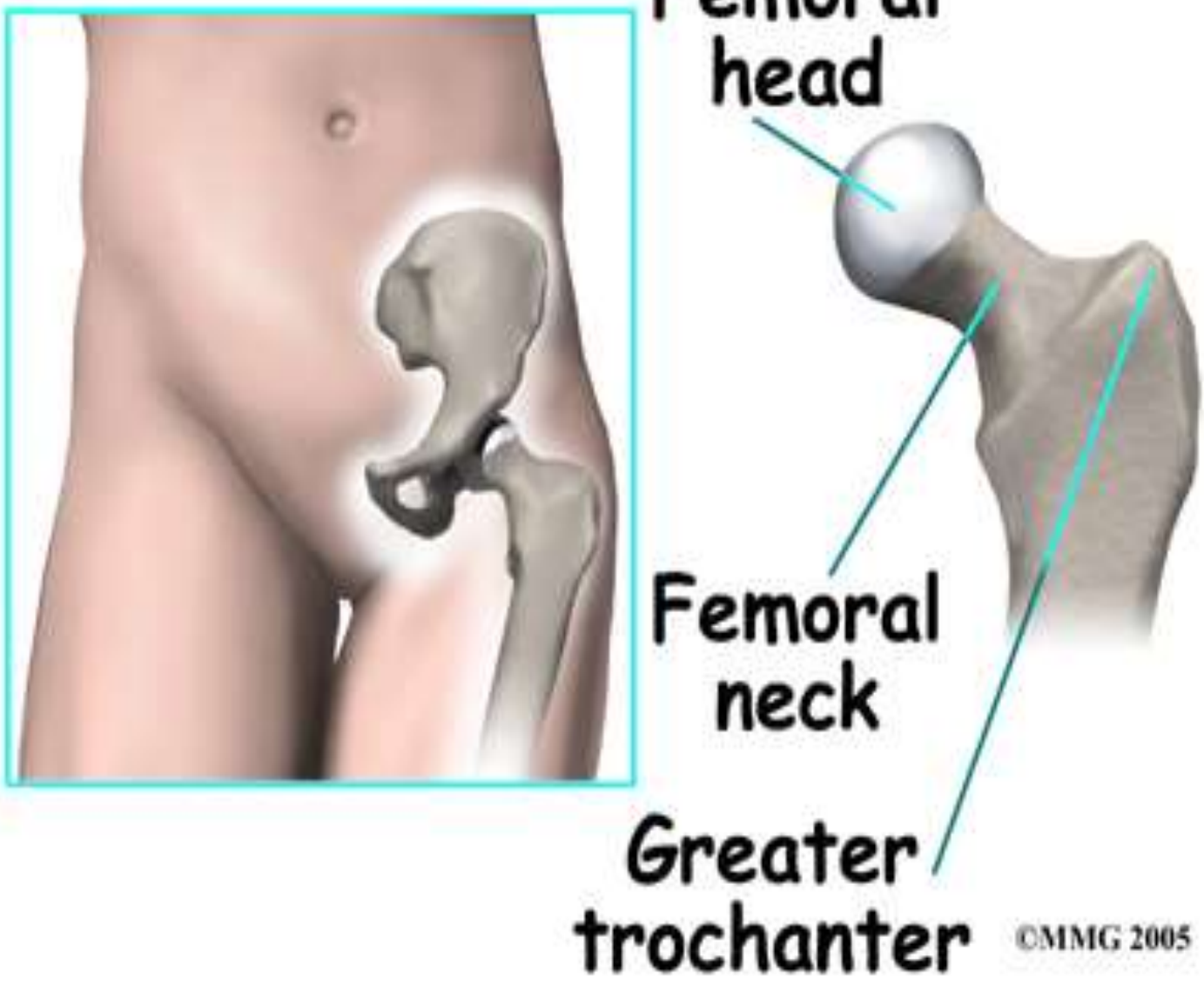
NURSING DIAGNOSIS

complication

Developmental Dysplasia of the Hip


Normal
hip joint
Dysplastic
hip joint
©MMG 2005


Acetabulum
Pelvis
Femoral head
Femur
Femoral neck
©MMG 2005


Femoral head
Femoral neck
Greater trochanter
©MMG 2005

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

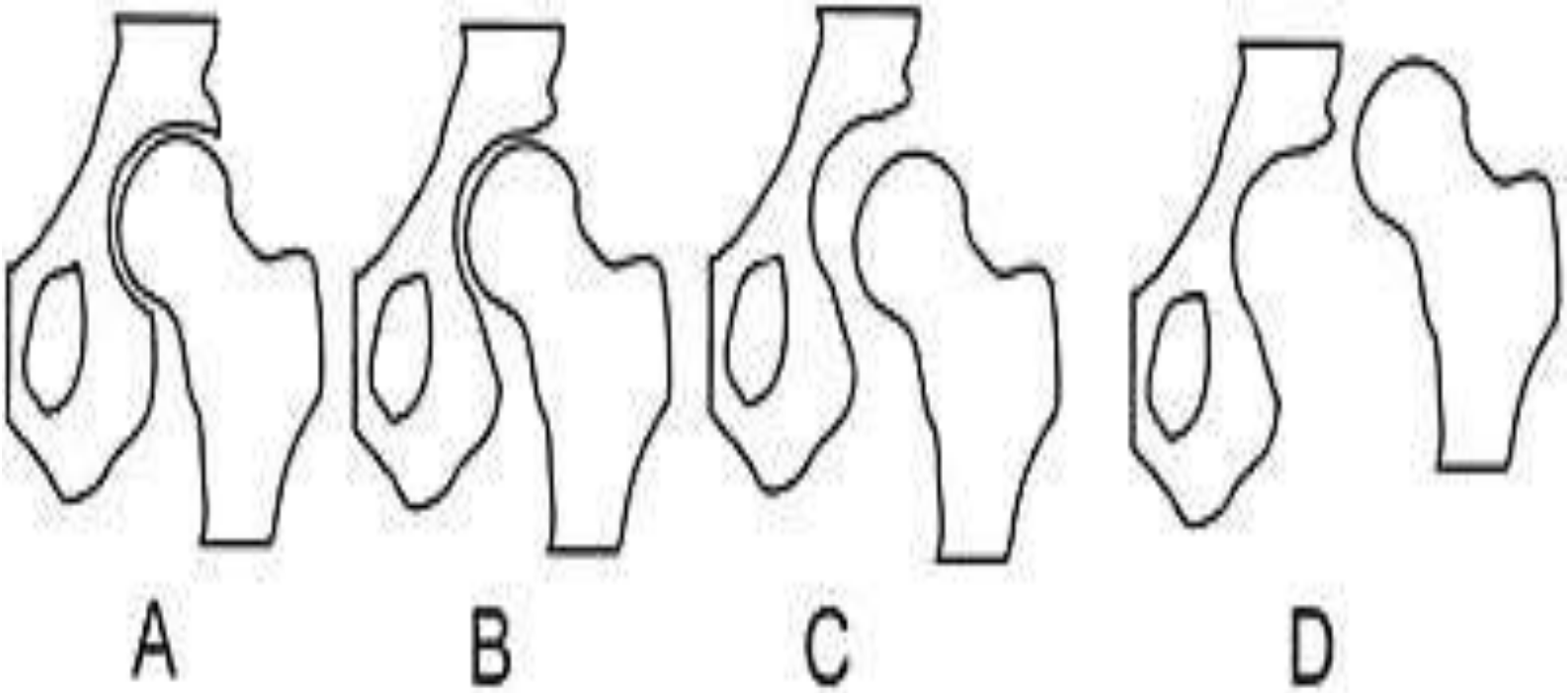
- i) Typical-neurological intact
- Ii) Terotological-involves neuromuscular defect such as arthrogyposis & myelodysplasia

- **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) **Dialocation**-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

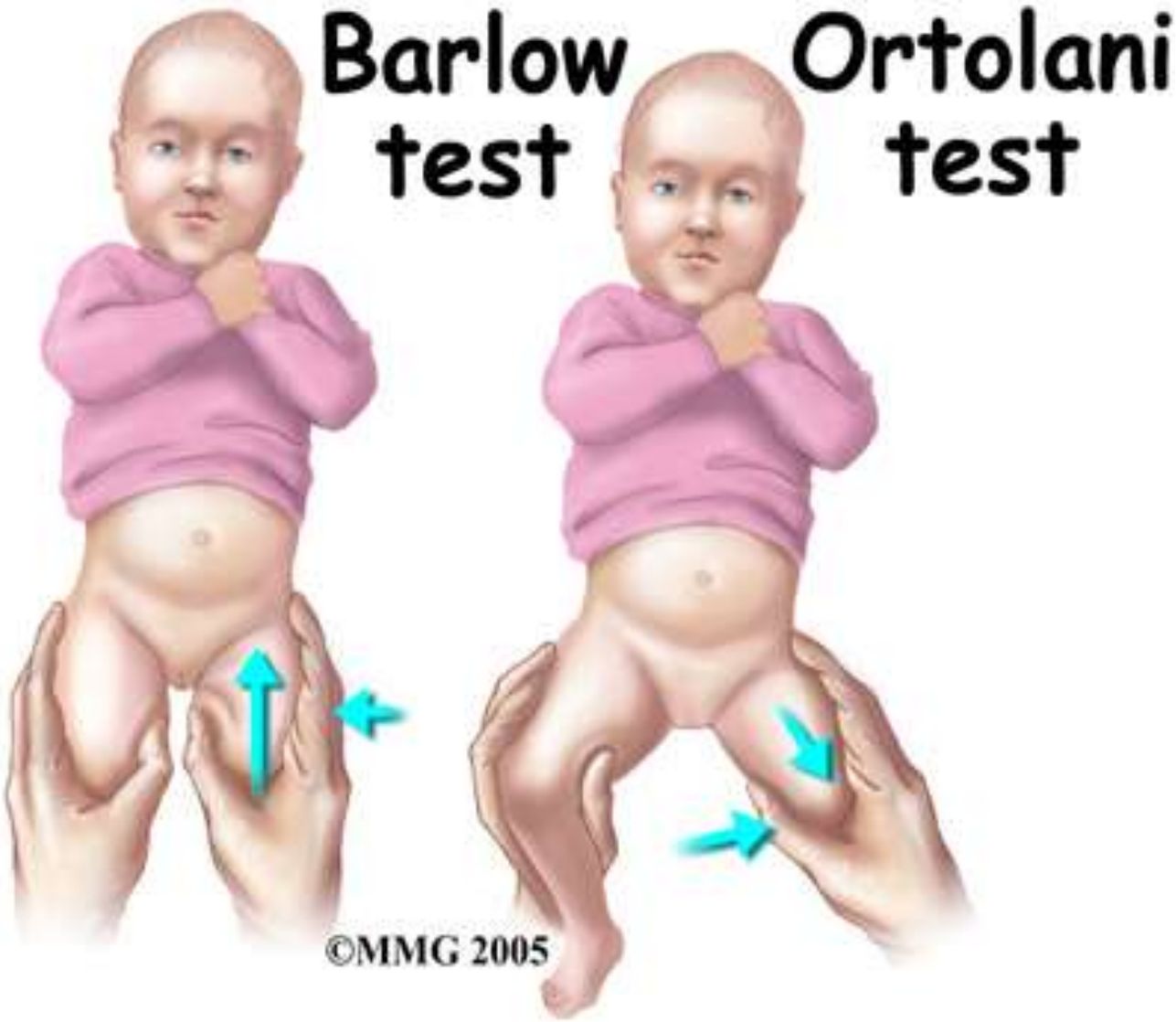
INFANTS

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
©MMG 2005

- 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
- Assymmetry 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

- *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

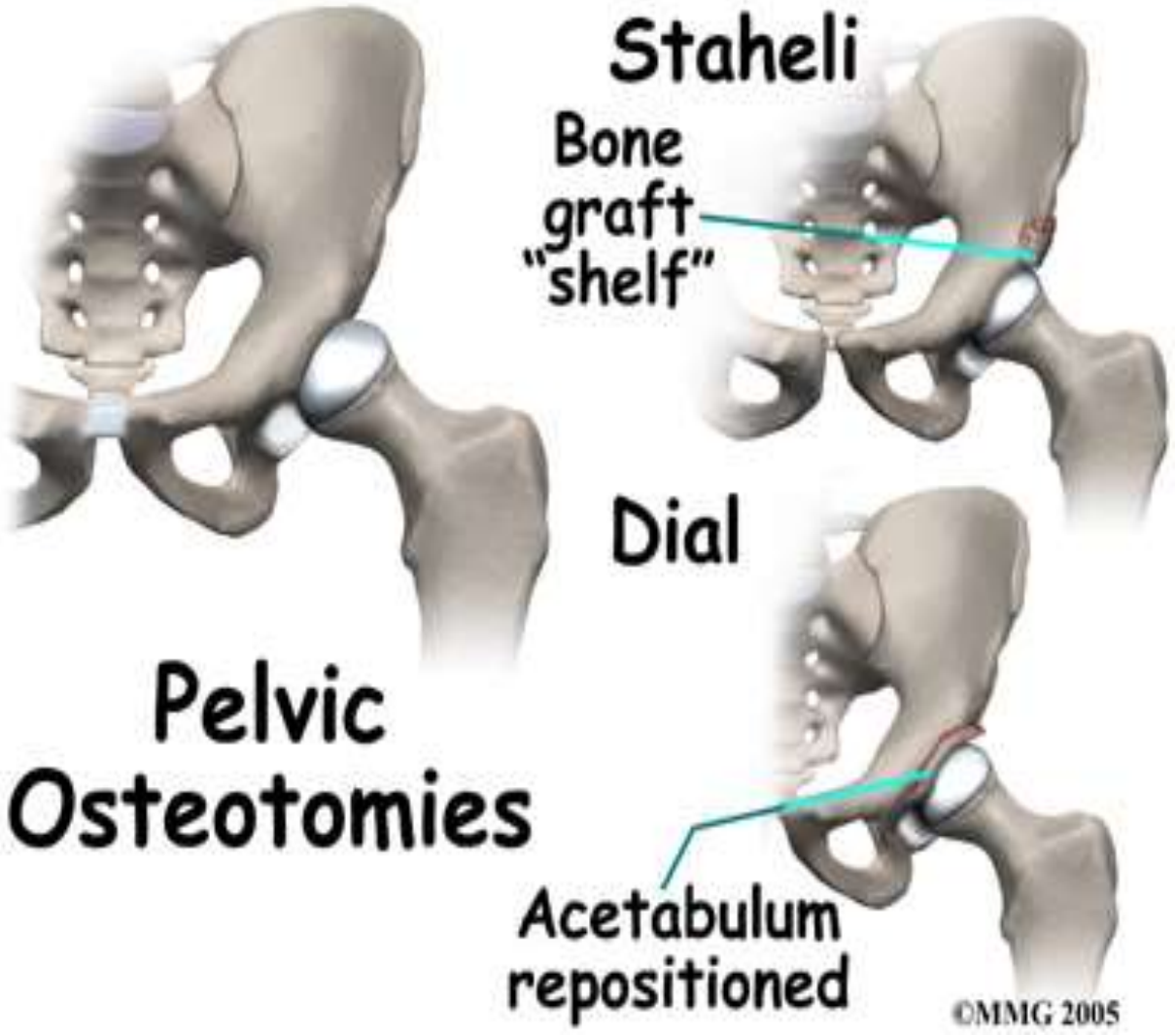
- . Traction consists of pulleys, strings, weights, and a metal frame attached over or on the bed.
- The purpose of traction is to stretch the soft tissues around the hip and to allow the femoral head to move back into the hip socket.
- Traction is most often used for approximately 10 to 14 days

Later infancy/toddler period

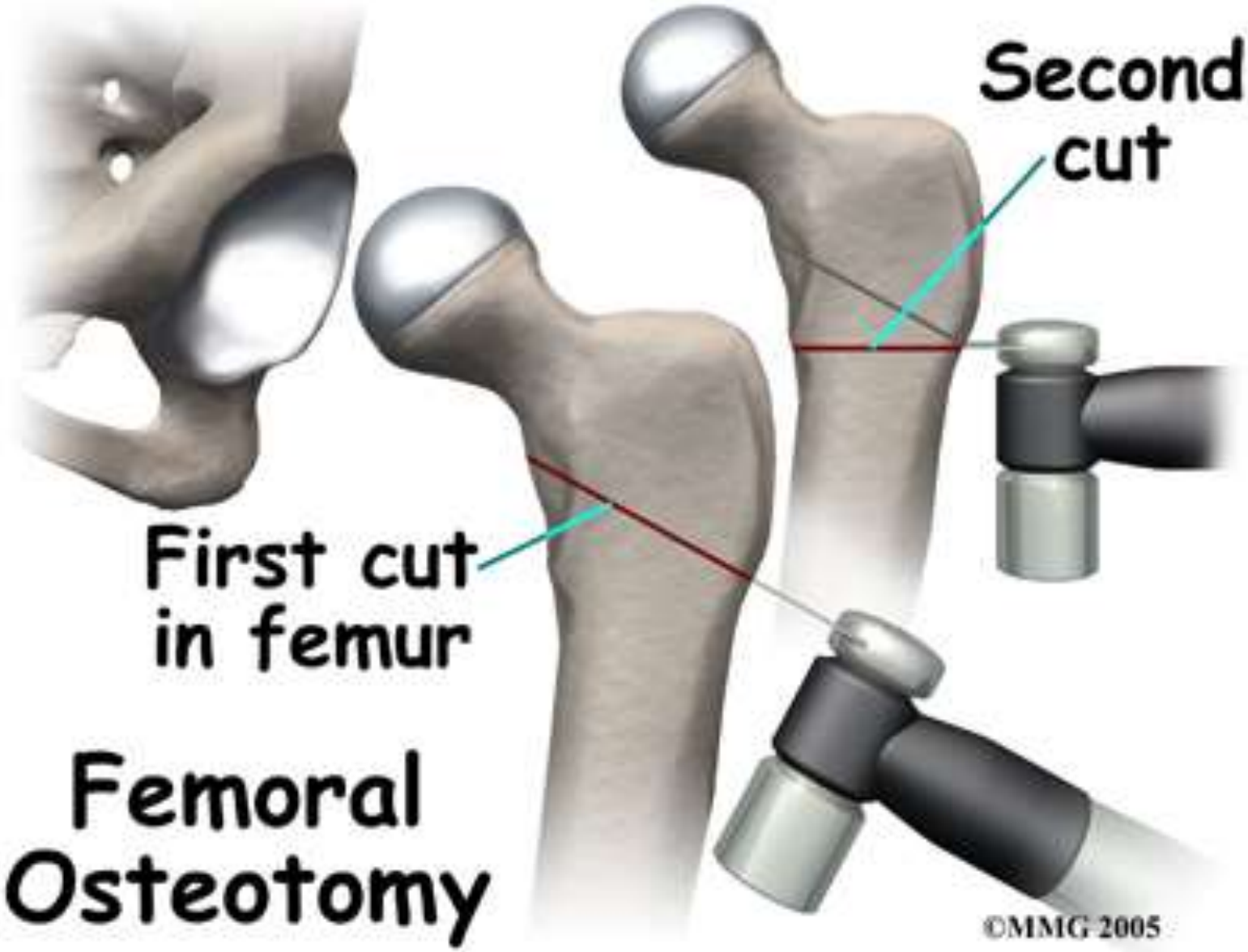
- Traction
- Closed reduction and open reduction of the hip
- Spica cast

Older children

- Preoperative traction
- Tonotomy
- Osteotomy



Pelvic Osteotomies


Second cut
First cut in femur
Femoral Osteotomy
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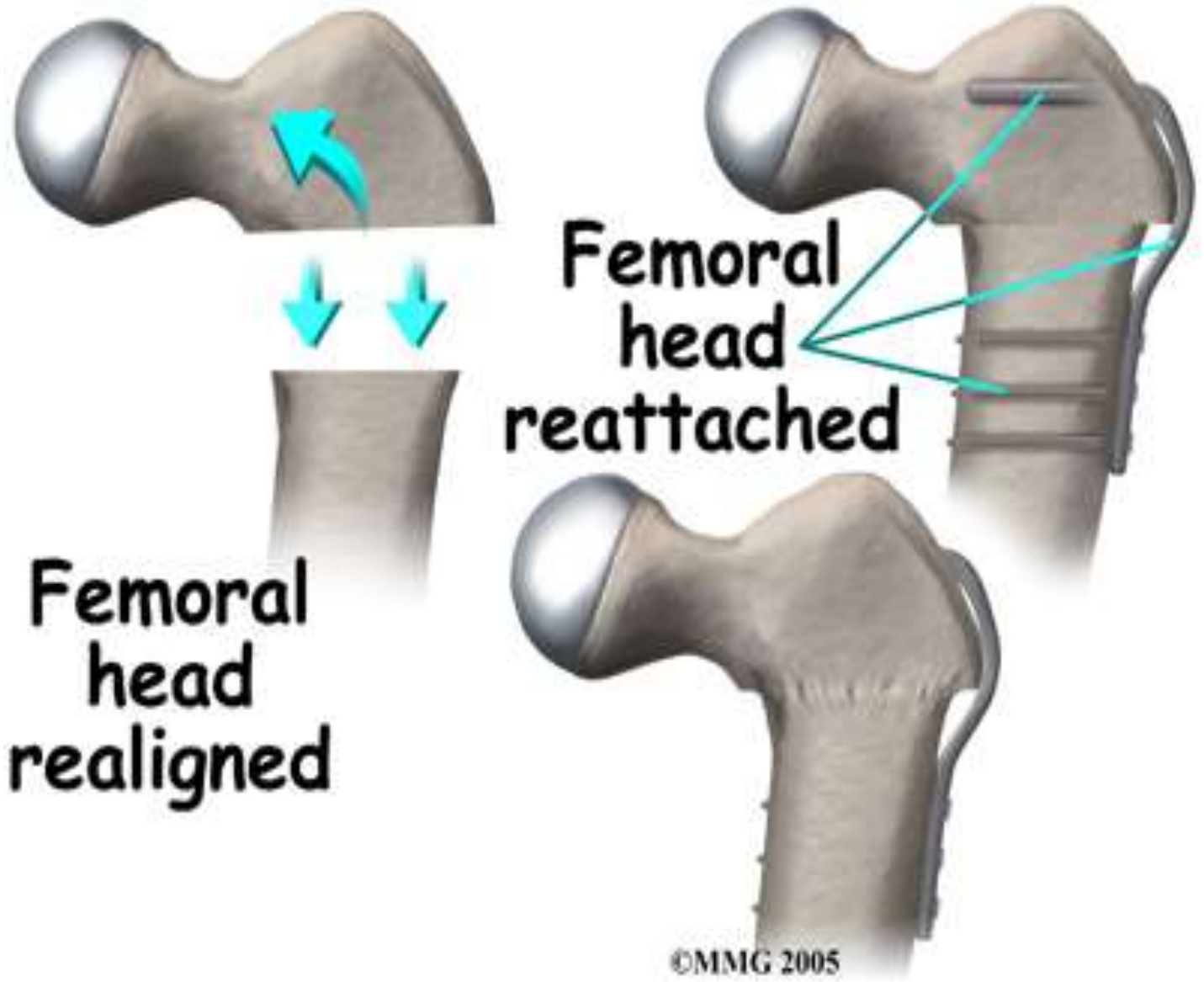
Femoral
head
reattached
Femoral
head
realigned
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NURSES RESPONSIBILITY

- 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

- A vascular 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