Pediatric and congenital Spine disorders
Dr. T. Myles

Mahmoud Benour R1

Objectives :

.Embryology Of the spine.

.Congenital Spine anomalies.

.Neural Tube Defect.

.Idiopathic Scoliosis.

Embryology of the spine

- **The spine is a complex and vital structure.**
- **Its function includes not only structural support of the body as a whole, but it also serves as a conduit for safe passage of the neural elements while allowing proper interaction with the brain.**
- **Anatomically, a variety of tissue types are represented in the spine.**
- **Embryologically, a detailed cascade of events must occur to result in the proper formation of both the musculoskeletal and neural elements of the spine.**

Embryology of the spine

- Alterations in these embryologic steps can result in one or more congenital abnormalities of the spine.
- Other body systems forming at the same time embryologically can be affected as well,resulting in associated defects in the cardiopulmonary system and the gastrointestinal and genitourinary tracts.

Embryology of the spine

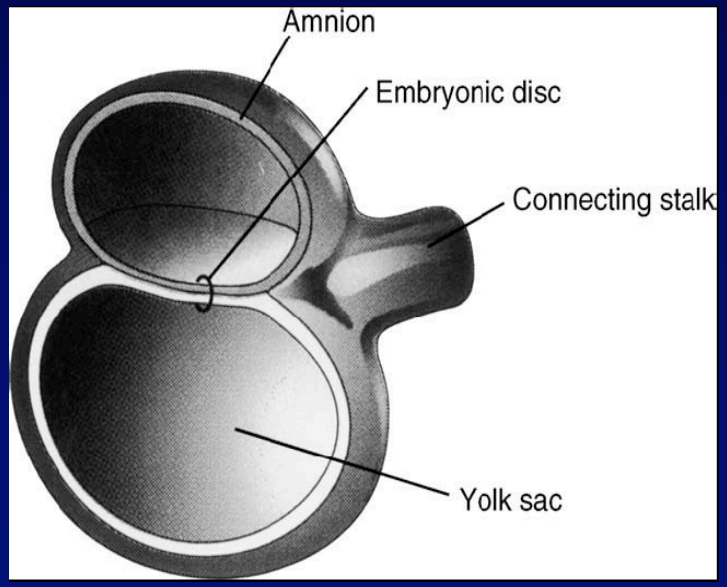
- Embryology of the spine involves two elements :

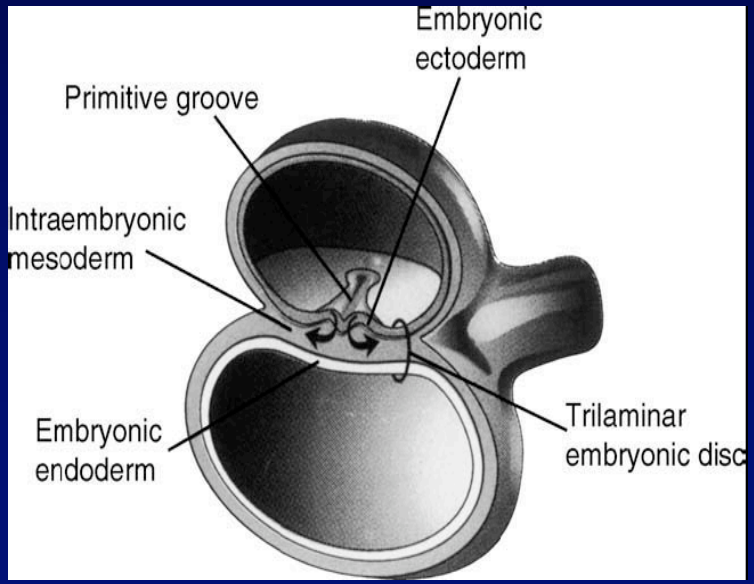
1. Musculoskeletal part.
2. Neural part.

Embryology of the spine

Musculoskeletal part :

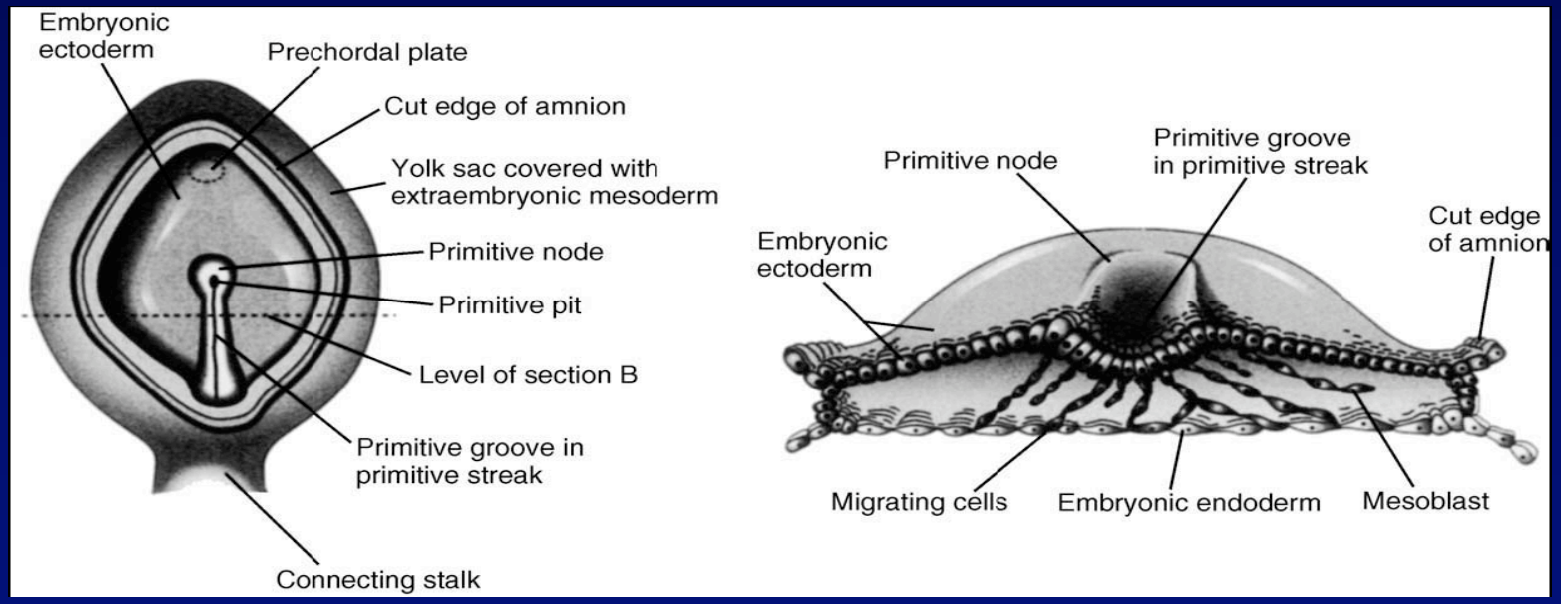
- The primitive streak, well defined germ layers and the notochord develop during gastrulation, which usually occurs during the third week of gestation.





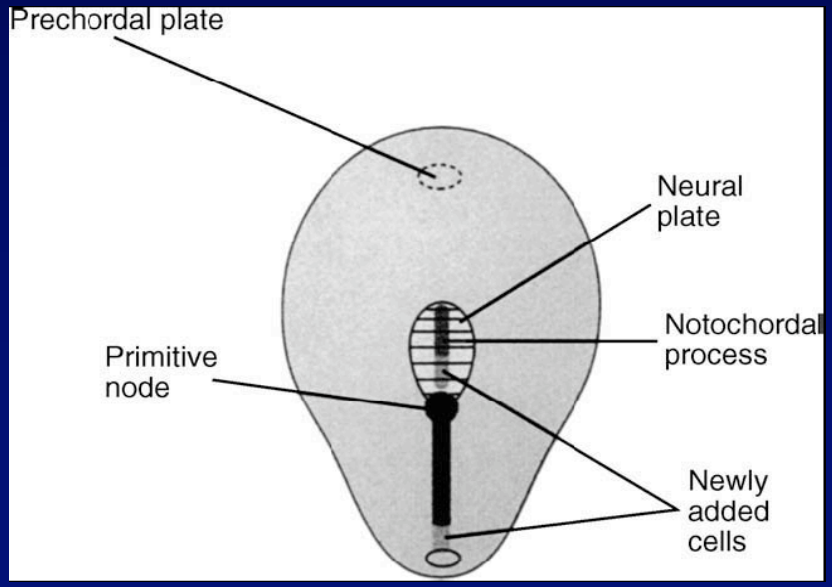
Embryology of the spine

- Epiblastic cells migrate from the deep surface of the primitive streak and form the embryonic endoderm.
- Subsequently, cells continue to migrate from the primitive streak, creating the embryonic mesoderm. The cells that remain on the epiblastic side of the embryonic disc form the embryonic ectoderm.



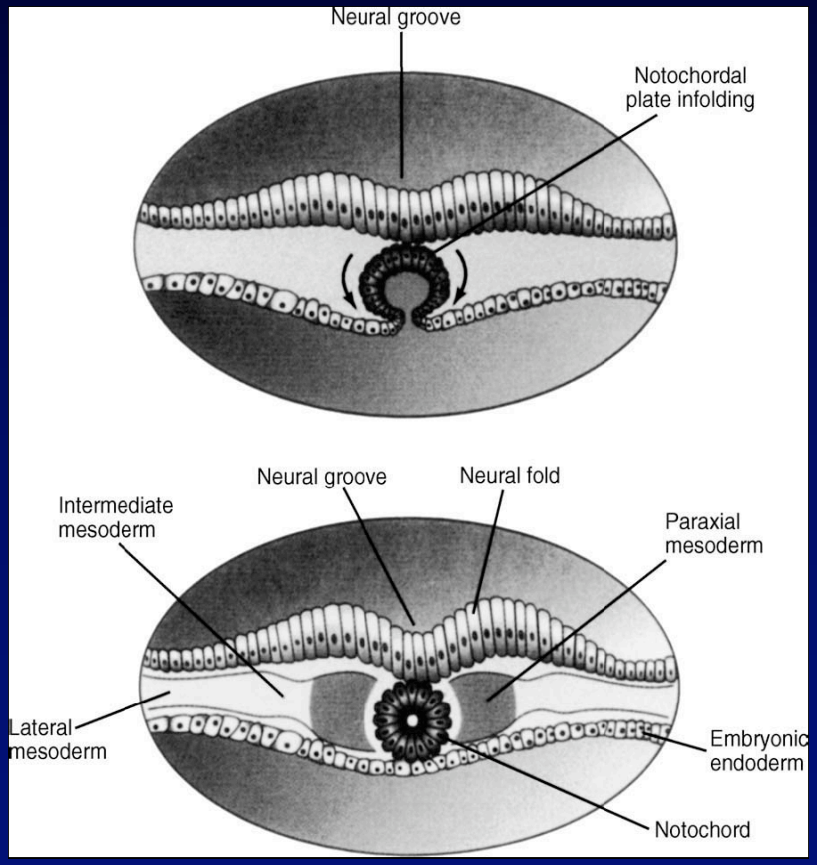
Embryology of the spine

- Cell migration continues into the fourth week of development after which the primitive streak regresses and disappears in the sacrococcygeal region.
- A group of specialized cells that migrate through the primitive node, which is located at the cranial end of the primitive streak, gives rise to the prechordal plate and notochordal process .



Embryology of the spine

- Cells migrate in from all areas of the primitive node. However, the cells migrating most anteriorly form the prechordal plate, whereas the ones migrating most posteriorly form the notochordal process will subsequently fold to form the notochord containing a central canal
- On both sides of the notochord, the mesoderm differentiates into three main areas: paraxial, intermediate and lateral mesoderm.
- 42 to 44 pairs of somites will form from the paraxial mesoderm by the end of the fifth week.
- Development of the somites occurs in a craniocaudal fashion and wil eventually help in forming the bones of the head, vertebrae, other bony structures of the thorax and associated musculature.



Embryology of the spine

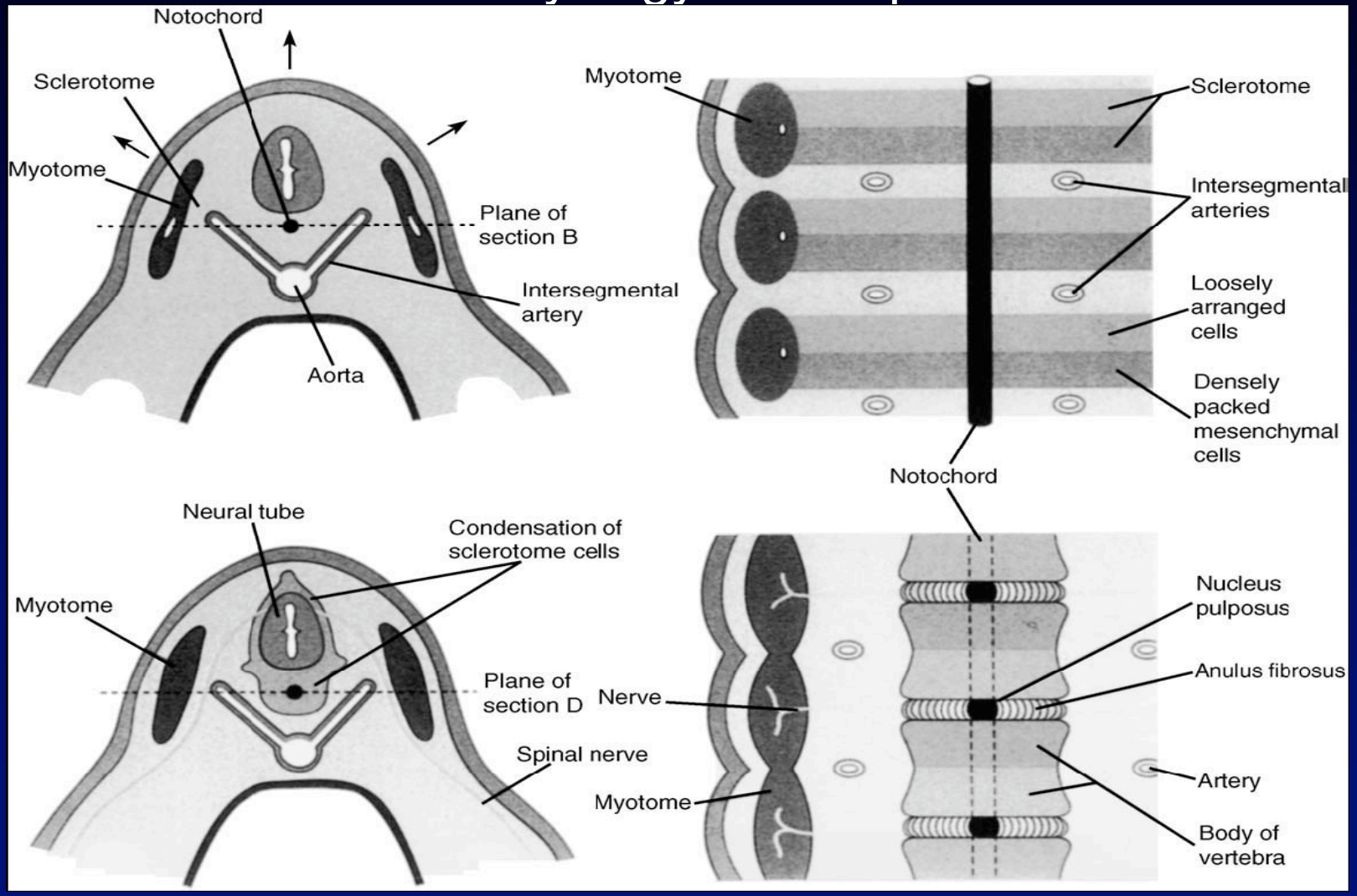
- Each somite develops into two parts:

1. A sclerotome → are responsible for the formation of the spine.
2. A dermomyotome → form muscle cells and the overlying dermis.

- During the fourth week, cells of the sclerotome begin to migrate toward and around the notochord and neural tube.
- Once the sclerotomes have surrounded the notochord and neural tube, each level will separate into :
    1. cranial area of loosely packed cells
    2. and caudal area of densely packed cells.

  The intervertebral disc will form between these two layers of cells.
- The area between the two halves in one level of the sclerotome is a “cell-free space.”

Embryology of the spine



Embryology of the spine

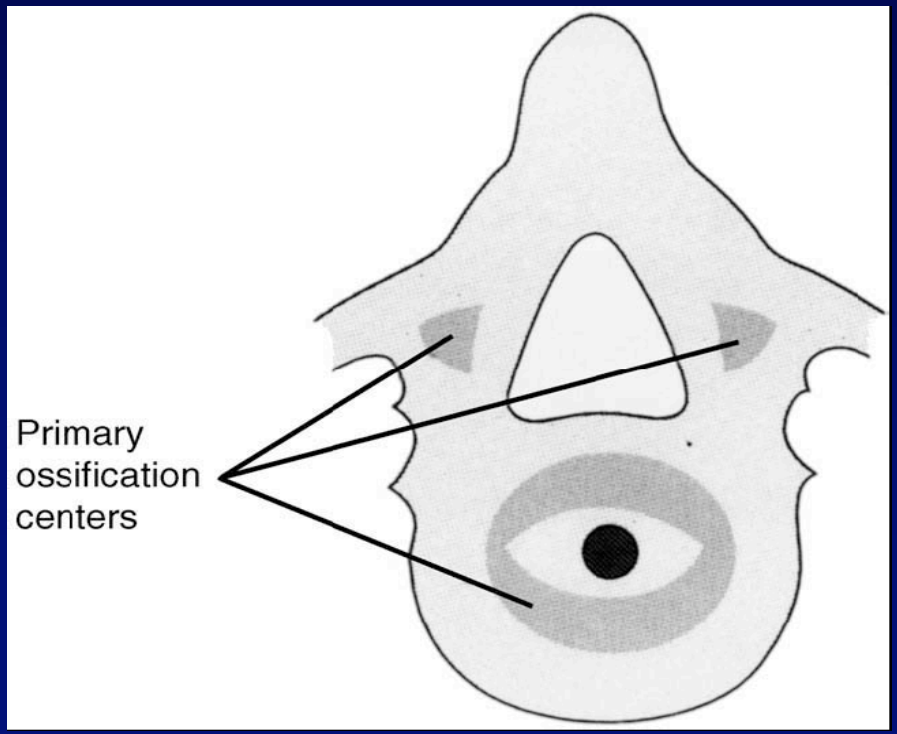
- The spinal nerve is associated with the caudal area of the sclerotome, and
  the intersegmental artery is located either between the somites or directly
  apposing the caudal area.
- The “cell-free space” will fill with cells migrating cranially from the caudal densely packed sclerotome layer to form the annulus fibrosus.
- The nucleus pulposus will develop inside the annulus from notochord.
- The developing intervertebral disc divides each sclerotome level and forces the remaining cells from a given densely packed layer to fuse with the loosely packed cells of the adjacent caudal level.
- As a result, one complete vertebra requires two somites to interact properly with each other in order to develop normally.
- Failure of this proper segmentation may result in a congenital abnormality.

Embryology of the spine

- Fusion of parts of the adjacent sclerotomes creates the centrum, which further develops into the vertebral body.
- The cells that initially migrated adjacent to the neural tube, develop into the neural arches,
- Vertebral arches consist of two pedicles and left and right halves of the laminae.
- The other processes associated with the posterior vertebral arch include the spinous process, transverse processes and articular processes.
- The centrum and the two halves of the vertebral arches develop separately and must fuse to one another.
- During the sixth week, after cells have migrated and vertebral structures begin to fuse, signals from the notochord and neural tube induce the chondrification of the relevant structures.

Embryology of the spine

- After chondrification, ossification will begin.
- Ossification centers can be found in three main areas in the vertebrae:
  one in the centrum and.
  one on each side of the vertebral arch.



Embryology of the spine

- The centrum or vertebral body will articulate with the vertebral arch at birth, with fusion occurring between the ages of 5 and 8 years.
- The two pieces of the arch begin to fuse during the first year of life with complete fusion occurring by age six years.
- Moore describes the five secondary ossification centers that form after birth; one for the tip of each transverse process.

  one for the extremity of the spinous process.

  one for the upper surface of the body.

  one for the lower surface of the body.
- It is important to mention that bone ossified from the secondary centers will contribute to the formation of growth plates.

Embryology of the spine

- congenital defect may result from absence or asymmetry of growth plates.
- Defects in both chondrification and ossification cause most congenital abnormalities of the spine.

Embryology of the spine

Neural part :

- The nervous system of the embryo is derived from the surface ectoderm of the trilaminar disc and begins to develop during the third week.
- The neural groove will fold into a tube and represents the future central nervous system.
- the peripheral nervous system develops from neural crest cells that migrated during the folding process.
- The brain develops from the most cranial aspects of the neural tube, and the spinal cord develops from the remaining caudal areas.
- The pia and arachnoid mater are both neural crest in origin.
- The dura mater is mesenchymal in origin.

Embryology of the spine

- In an embryo, the spinal cord fills the canal, but as a result of normal growth and development, it will eventually change positions.
- After birth, as a child develops, the spinal cord will continue to ascend to the level of the first or second lumbar vertebra. The remaining caudal aspect of the spinal canal becomes a conduit for the lumbar and sacral nerve roots

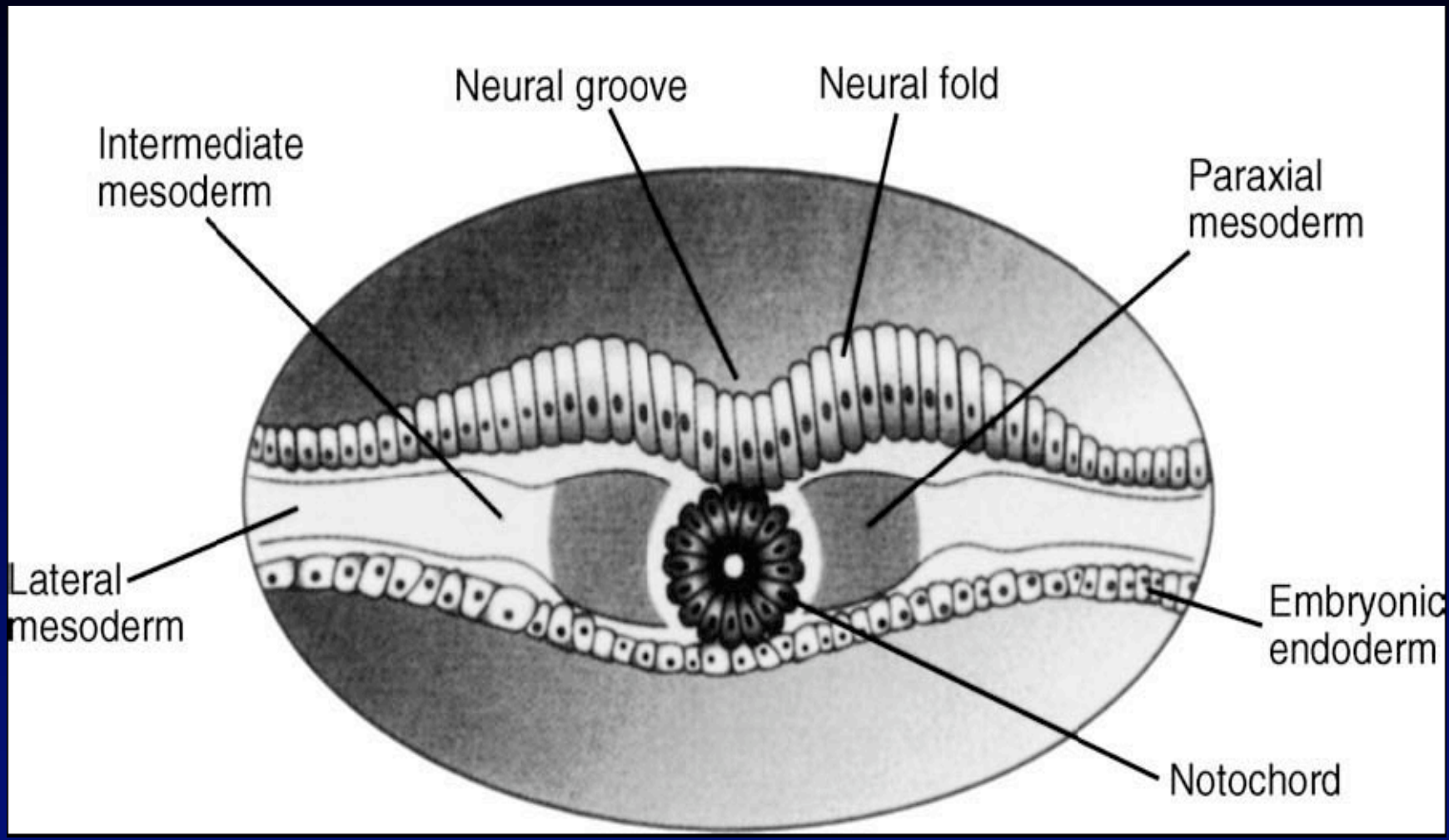
  (cauda equina).
- Nerve roots leave the spinal cord at angles as a result of the normal spinal cord ascension.
- During the fetal period, the normal curve of the vertebral column is kyphotic.
- Lordosis in the cervical vertebrae develops as a result of the infant holding its head upright. Lumbar lordosis similarly develops secondarily as a result of the infant achieving a sitting and then standing posture.

Development of additional organs and systems:

- Mesoderm has 3 parts :

1. Paraxial mesoderm.
2. Intermediate mesoderm.
3. Lateral mesoderm.

- Paraxial mesoderm is responsible for the formation of the vertebrae as well as the dermis of the skin, striated skeletal muscle, muscles of the head and connective tissue.
- In addition, the other two areas of mesoderm, intermediate and lateral, are involved in the development of the urogenital, pulmonary and cardiac systems.
- Thus, a defect affecting the development of the mesenchyme responsible for bony formation of the spine may also be responsible for defects in alternative organ systems.


Neural groove
Neural fold
Intermediate mesoderm
Paraxial mesoderm
Lateral mesoderm
Embryonic endoderm
Notochord

- The main associated defects involve the VACTERL syndrome.
- **VACTERL** is **V**ertebral anomalies, **I**mperforate anus, **C**ardiac abnormalities, **T**racheoesophageal fistula, **R**enal dysplasias and **L**imb malformations.
- Another classic syndrome associated with congenital spinal defects is Klippel-Feil.
- The genitourinary system may be the most frequently involved system with congenital abnormalities of the spine. Due to fact that the mesoderm forming the vertebrae is also responsible for the formation of the mesonephros, the predecessor of the mature genitourinary system.
- The medial region of this mesoderm forms the vertebrae while the ventrolateral region forms the mesonephros.

Classification of congenital abnormalities:

- It is important to identify :

1. The type of malformation.
2. The resulting deformity.
3. The specific region of the spine where the malformation occurs.

- Malformations of the spine can be classified into three main groups:

1. Neural tube defects.
2. Defects of segmentation.
3. Defects of formation.

- Neural tube defects

  a condition in which the neural tube fails to completely close during the fourth week of embryonic development. As a result, structures overlying

  these midline abnormalities are severely affected and may be unable to form.

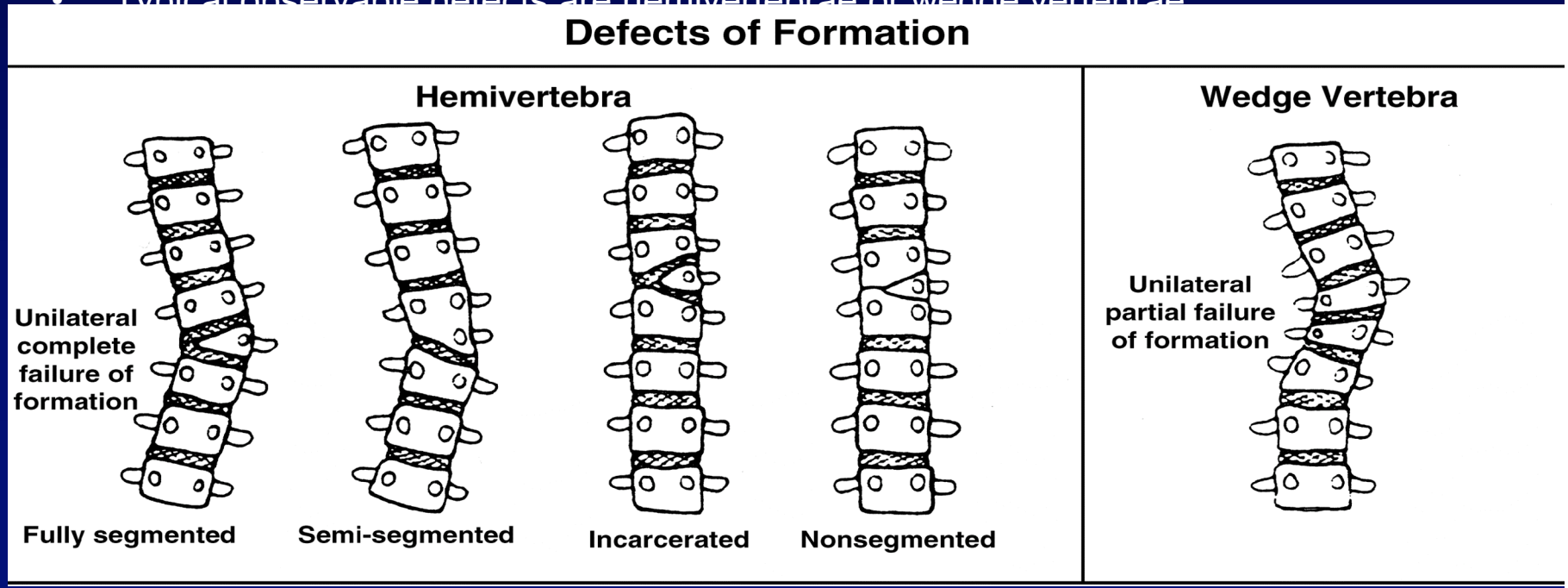
- Causes :

- Nutritional factors.
- Environmental factors.
- Drugs, such as anticonvulsants.

- In-utero diagnosis of a neural tube defect may be considered when there is an elevated level of alpha-fetoprotein.

Defects of formation and segmentation :

- Failures of formation arise as a result of an absence of a structural element of a vertebra. Any region of the vertebral ring may be affected: anterior, anterolateral, posterior, posterolateral and lateral.
- The type of deformity depends on the area of the vertebral ring affected, which will alter normal growth patterns.
- Typical observable defects are hemivertebrae or wedge vertebrae



Defects of formation and segmentation :

- Hemivertebrae are bony remnants that did not complete normal development and can be :

1. **Fully segmented**. still have growth plates both cranially and caudally.

2. **Semisegmented.** fusion with a cranial or caudal vertebra.

Thus, in a semisegmented vertebra, there is a functional disc on one side only.

3. **Nonsegmented.** has not separated from either the cranial or caudal vertebra.

Defects of formation and segmentation :

- **Incarcerated hemivertebrae** :

  When both the cranial and caudal vertebrae conform in shape to make room for the hemivertebra, the anomalous vertebra is referred to as incarcerated.

  Incarcerated hemivertebrae may not affect the shape of the spine. The pedicles of an incarcerated hemivertebra are in line with the curve created by the pedicle cranial and caudal to it.

- **Nonincarcerated hemivertebra** is always fully segmented, and the potential wedge effect of these anomalies results in a higher likelihood of progression. In this case, the pedicles are outside of the pedicle line of the adjacent vertebrae.

Defects of segmentation :

- Failures of segmentation occur when two or more vertebrae fail to fully separate and divide with concomitant partial or complete loss of a growth plate.
- These defects are classified depending on the region and quantity of vertebrae affected :
- **Block vertebrae** Involvement of entire vertebrae.

- **Unilateral bars** Involvement of specific regions of the vertebral ring create bar that act as an asymmetric rigid tether to normal growth.

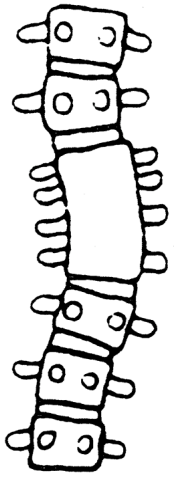
- **Unilateral unsegmented bars with a contralateral hemivertebra**.

Defects of segmentation :

Defects of Segmentation

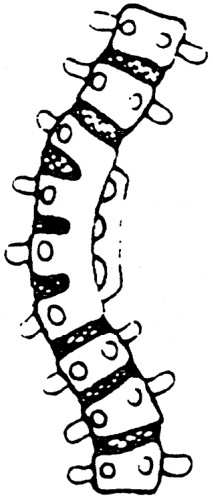
Block Vertebra

Bilateral failure of segmentation

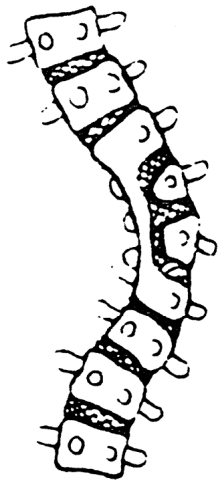


Unilateral Bar

Unilateral failure of segmentation



Unilateral Bar & Hemivertebra



Spina bifida oculta :

- Results from the failure of fusion or development of part of the vertebral arch, usually lamina.
- Does not involve the spinal cord or meninges.
- Approximately 10% to 24% of population.
- usually asymptomatic, may presented with a skin indentation and a patch of hair growing in the area of the lesion.
- X-ray examination is the only valid test to confirm this type of neural tube defect.
- If the patient shows no associated abnormalities, no further treatment is necessary.

Spina bifida cystica :

- **Spina bifida with meningocele :** When the cyst involves the meninges and cerebrospinal fluid.

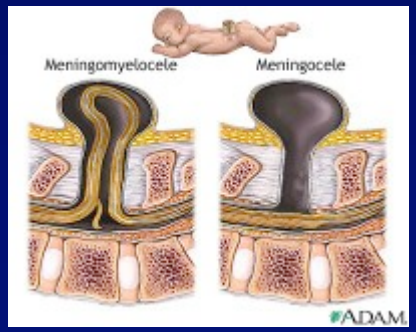
  Usually have a layer of normal epidermis covering the meninges.

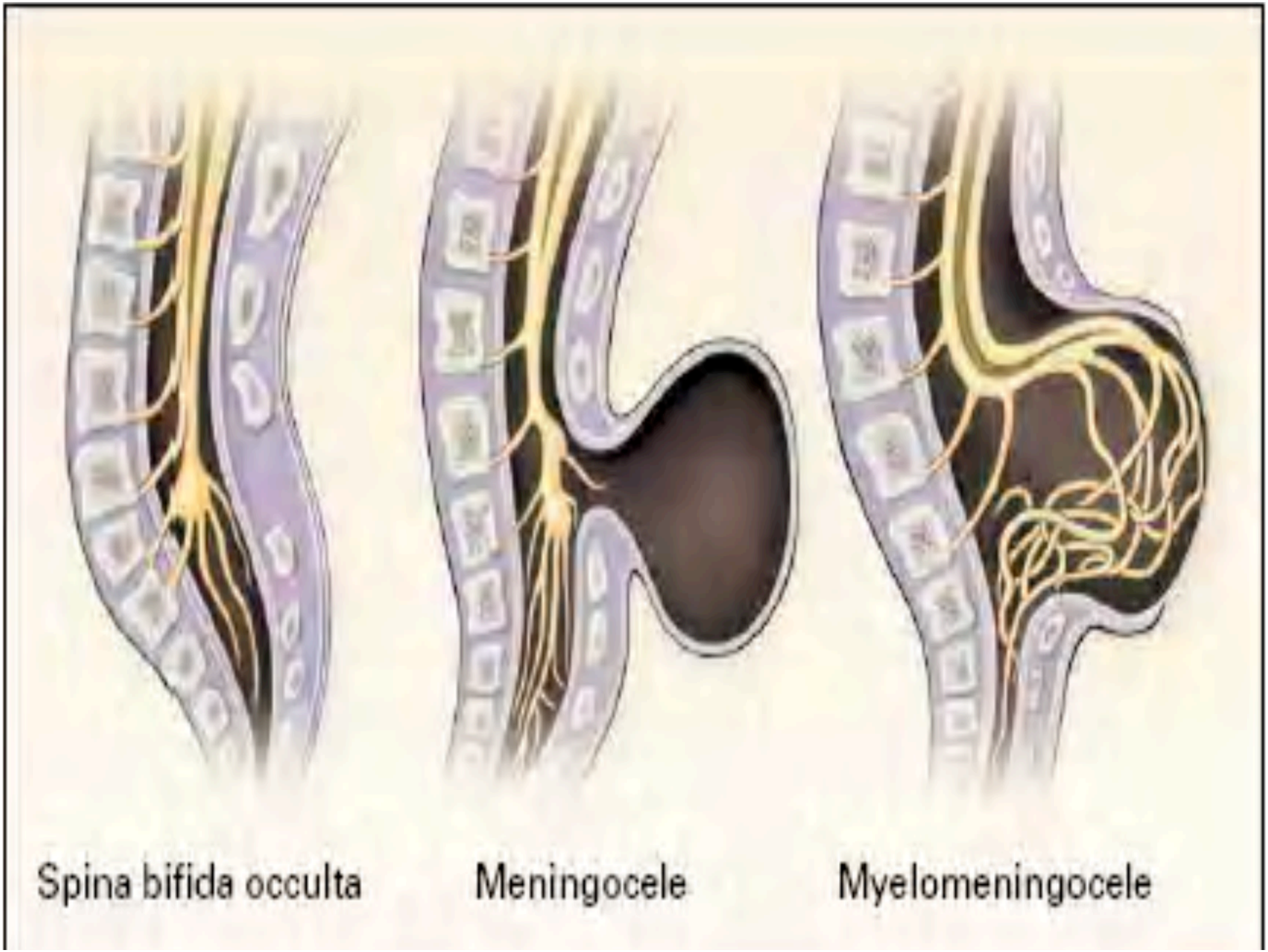
  External manifestations that may indicate this type of neural tube defect include hair growth in the area of the lesion, lipomas, cysts or hemangiomas
- **spina bifida with meningomyelocele :** When the cyst involves the meninges, cerebrospinal fluid,and also contains the spinal cord.

  More common than spina bifida with meningocele.

  It is considered a more severe neural tube defect because of the involvement of the spinal cord and meninges.

  May be covered by a thin layer of skin or by a membranous sac.




Spina bifida occulta
Meningocele
Myelomeningocele

Spina bifida with meningomyelocele :

- Neurological symptoms such as :

  limb paralysis.

  bladder and bowel incontinence.

  hip dislocations.
- Because this defect may involve just the nerve roots or the entire spinal cord, paralysis may be of the flaccid, spastic or mixed type.
- Other associated abnormalities may include :

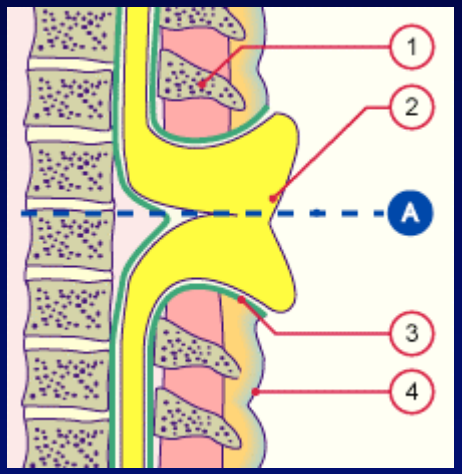
  Hydrocephalus,

  Arnold-Chiari malformations and

  Severe forms of scoliosis, kyphosis or lordosis.

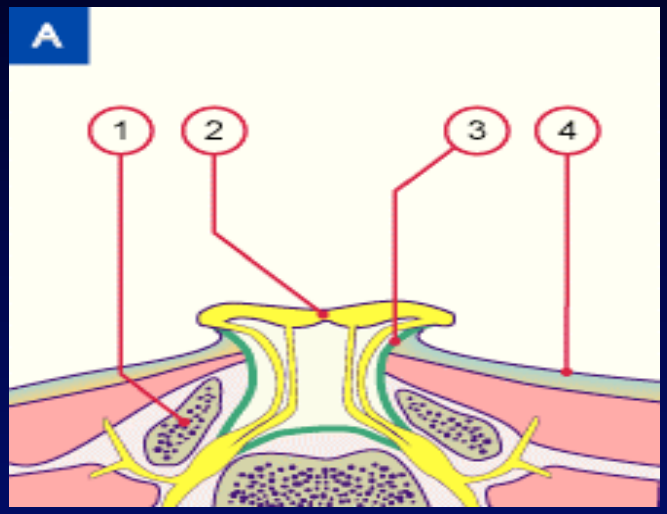
Myeloschisis :

- The most severe type of spina bifida. This defect occurs as a result of the neural tube not closing properly with the developed spinal cord being exposed to the external environment.



1.Spinous process
2.Spinal cord
3.Dura mater
4.Skin up to the neural tube defect

Myeloschisis :



1.Vertebral arch
2.Spinal cord
3.Dura mater
4.Skin up to the neural tube defect

Diastematomyelia :

- Definition:

  Longitudinal clefting of the spinal cord that is divided into two hemi-cords

- Pathology:

  The spinal cord and the nerve roots are splitted into two columns.

  It may occur with spina bifida or with a closed spine.

  It could be bony, fibrous or cartilaginous septum that subdivides partially or completely the vertebral canal.

  The hemicords usually have a distinct arachnoid membrane each with a common dura.

  It may involve single vertebra or extend to several vertebral segments.

  The cleft may be found at any level, but in most cases is found at the lower thoracic or upper lumbar regions.

Associated anomalies :

1) Open spina bifida.

2) Closed abnormalities of the vertebrae: scoliosis, kiphosis, hemivertebrae, butterfly vertebra.

3) Cutaneous manifestations on the dorsal midline consisting of telangiectasias, atrophic skin, hemangiomas, subcutaneous lipomas and cutaneous nevi. Among the cutaneous nevi, the most characteristic is the nevus pilosus, a large patch of long silky hairs, that is situated over the site of the cleft in the cord in 50-70% of the cases. The location of the cutaneous abnormality, however, is necessarily indicative of the level of the lesion.

4) Orthopedic deformities of the feet, especially clubfoot, are found in approximately half of the patients.

Pathogenesis :

- **Several hypothesis have been formulated:**

1) retention of the neurenteric canal that transiently connects the yolk sac to the amnion via the primitive knot. This knot migrates distally to the region of the coccyx, where it disappears. If an accessory canal develops, it would split the neural ectoderm with underlying endoderm and result in a midline fistula. The fistula eventually disappears, but not until abnormal vertebral and neural elements have been formed

2) presence of a dorsal and ventral cleft that severs the neural plate near the midline, resulting in separate closure of the two hemicords. Mesenchymal tissue filling the gap results in mesodermal and bony abnormalities.

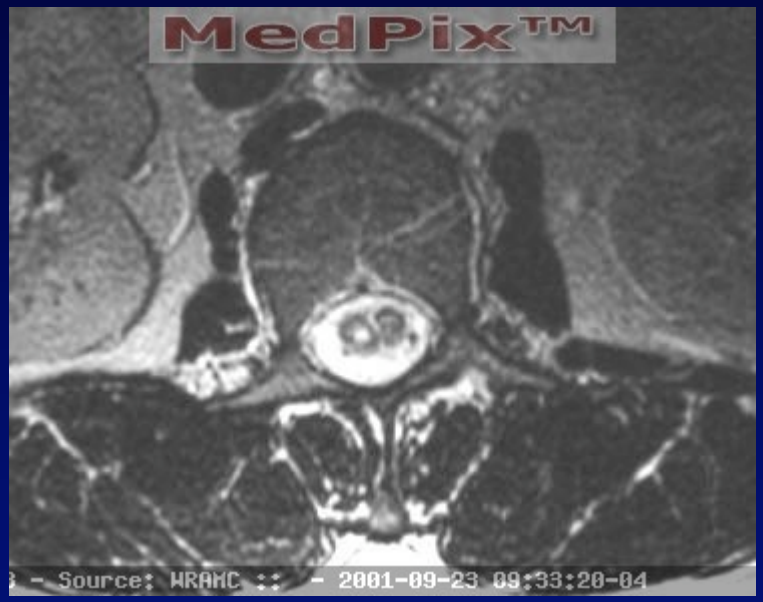
3) excessive dilatation of the neural tube resulting in subdivision of the cord and internal penetration by mesodermal structures originating from the vertebral body.

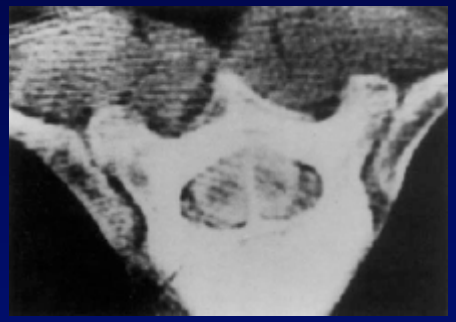
- Diastematomyelia acts as a restraint that slows the normal growth of the spinal cord by impeding the upward migration of the neural elements, with progressive neurologic deficits in the limbs.

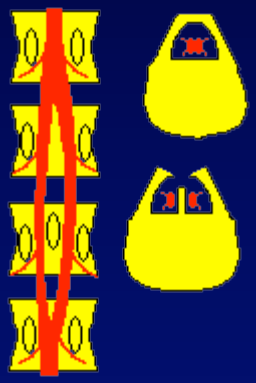
- Etiology:

  Unknown.

Diastematomyelia







Congenital scoliosis

- An abnormal vertebral development that results in a lateral curvature of the spine is classified as congenital scoliosis.
- Due to failure of formation or a failure of segmentation, combined defects are the most common in congenital scoliosis.
- In addition, a sagittal deformity can be involved, creating either a kyphoscoliosis or a lordoscoliosis.
- 64% of cases of congenital scoliosis involve the thoracic vertebrae.

  20% involve the thoracolumbar region.

  11% of the cases are seen in the lumbar region.

  5% in the lumbosacral region.

  (According to Jaskwhich et al.)
- Although unusual, congenital scoliosis does occur in the cervical or cervicothoracic regions as well

Congenital scoliosis :

- majority of curves are progressive, whereas only 25% are nonprogressive.
- Curves involving the thoracic vertebrae show the poorest prognosis.
- Curves in the cervicothoracic and lumbosacral regions are severe because there is a diminished ability for compensation by the rest of the spine.

- The prognosis of vertebral abnormalities in order beginning with poorest:
  unilateral unsegmented bar.
  Double-convex hemivertebrae.
  Single free convex hemivertebra.
  Block vertebrae.

- Anomalies may be associated with congenital scoliosis :
  The genitourinary tract, cardiac system and the spinal cord are the most common areas for associated abnormalities in scoliosis.
- Special consideration should be given to abnormalities associated with the spinal cord, because they may require surgical treatment.

NATURAL HISTORY

- The progression of congenital scoliosis depends on both the type and the location of the vertebral anomaly. Curve progression is caused by unbalanced growth of 1 side of the spine relative to the other.

- The best natural history study to date is that of McMaster and Ohtsuka, who reported that the rate of deterioration and severity of final deformity were predictable according to the type of anomaly and location of the curve.
- They found that only 11% of cases were nonprogressive, whereas 14% were slightly progressive, and the remaining 75% progressed significantly.

NATURAL HISTORY

- The prognosis for congenital scoliosis with regard to its rate of deterioration and final severity depends on many factors, as follows:

1. **Type of vertebral anomaly:**

Risk of Progression (Highest to Lowest)	**Curve Progression**
Unilateral unsegmented bar with contralateral hemivertebra	Rapid and relentless
Unilateral unsegmented bar	Rapid
Fully segmented hemivertebra	Steady
Partial segmented hemivertebra	Less rapid
Incarcerated hemivertebra	May slowly progress
Nonsegmented hemivertebra	Little progression

NATURAL HISTORY

2.Site of the anomaly:

.Most severe in the thoracic and thoracolumbar regions.

.Less severe in the cervicothoracic and lumbar regions.

3. Age of the patient at the time of diagnosis:

.First few years of life has a particularly bad prognosis.

ASSOCIATED ANOMALIES

- Neural axis abnormalities are present in up to 35% of patients.
  Diastematomyelia (split cord).
  Cord tethering.
  Chiari malformations.
  Intradural lipomas.
- Congenital heart disease is observed in up to 25% of patients.

- Genitourinary anomalies are observed in up to 20% of patients.
- Musculoskeletal anomalies.
  Clubfeet.
  Sprengel deformity. (elevation & medial rotation of inferior scapula).
  Developmental dysplasia of the hip.

Clinical evaluation

- History Taking:

  .Includes Maternal perinatal history, family history, and developmental milestones must be fully explored.

- Full physical examination

  .Includes evaluation for hearing, visual, and dental problems; cleft palate and cleft lip; hernias, anorectal abnormalities, and genitourinary problems, cardiac murmurs and neurologic disorders.

  .The spine is best examined with the patient fully disrobed and erect.

  .Skin dimples and hair patches should be documented.

  .Shoulder and pelvic levelness are assessed, and asymmetry and prominence of the scapulae are noted.

  .With the patient forward-flexed, rotational deformity is recorded as centimeters of rib hump, and any lack of spinal motion is documented.

Radiographic evaluation

- Standard posteroanterior (PA) and lateral views of the entire spine are used for the initial evaluation.
- The quality of the bone and disk spaces on the convexity must be clearly visualized and inspected. If the disk spaces are present and clearly defined and the convex pedicles clearly formed, convex growth is possible, and the prognosis is poor.

- CT and MRI are useful in detailing bony canal anatomy and associated spinal cord abnormalities. These evaluations are mandatory prior to surgical intervention because spinal cord tethering and diastematomyelia must be identified and released prior to correction of the curve.

Treatment

- The primary goal of treatment of congenital scoliosis is to prevent the development of a severe deformity.
- Three key factors exist in achieving an optimum result in patients with congenital scoliosis, as follows:

1.Early diagnosis

2.Anticipation: based on

The amount of spinal growth remaining.

The type and site of the vertebral anomaly.

3.Prevention of deterioration: It is easier to prevent a severe spinal deformity

than to correct. All patients require radiological assessment at

4 to 6 month intervals, and once progression is established, immediate treatment is necessary to prevent further deterioration.

Nonsurgical management

- Observation, monitoring the curves for progression.
- Radiographs are obtained at regular visits (every 4-6 mo). The same vertebrae are measured on each radiograph, using exactly the same vertebral landmarks, and the current film is compared to the film from the last visit and the original film.
- The two periods of rapid growth are during the first 4 years of life and during adolescence. More frequent visits are necessary during these periods.
- Bracing is contraindicated in : short stiff curves, an unsegmented bar, congenital lordosis, and congenital kyphosis. (Natural history indicates a poor prognosis).
- Braces are unlikely to be effective if the scoliosis is more than 40° or if less than 50% flexibility is established using side bending or distraction radiographs.

Nonsurgical management

- For high thoracic curves the brace of choice is the Milwaukee brace.
- TLSO for lower thoracic curves.
- After surgery, however, a brace may be required to help control spinal alignment and the development of compensatory curves that were not included in the fusion.

Operative treatment

- The method of surgery selected depends on :

1. The age of the patient.
2. The site and type of vertebral anomaly.
3. The size of the curvature.
4. The presence of other congenital anomalies.

- Successful surgical treatment depends on selecting the right procedure and applying it at the right time.
- There are 4 basic procedures for the surgical treatment of congenital scoliosis : 1.Convex growth arrest (anterior and posterior hemiepiphysiodesis).

2.Posterior fusion.

3. Combined anterior and posterior fusion.

4. Hemivertebra excision.

Convex growth arrest

- First described by MacLennan in 1922.
- It was designed to arrest the excessive convex growth and allow the concave growth to occur and correct the deformity.
- Relatively safe procedure.
- The surgery is performed in 2 stages that usually are carried out under the same anesthetic. The spine is first approached anteriorly on the convexity of the scoliosis. The lateral half of the disks and their adjacent endplates are removed at the site of hemivertebra and at one intervertebral level above and below. This removes the anterior growth plates at the site of the anomaly, which is the main cause of the increasing scoliosis.
- The second stage of the procedure is performed through the separate posterior exposure of the convexity of the curve at the site of the hemivertebra. The paraspinal muscles on the concavity of the curve should not be stripped. A posterior convex fusion is performed to one level above and below the hemivertebra.

Posterior fusion

- The aim of a posterior fusion is not curve correction, but rather curve stabilization with the prevention of further curve increase.
- The fusion must cover the entire measured curve.
- Posterior spinal instrumentation achieves a moderately better correction and reduced incidence of pseudoarthrosis compared with a posterior spinal fusion.
- Spinal instrumentation is, however, associated with a greater risk of producing neurologic complications due to the effect of distraction on the spinal cord while the patient is anesthetized.

Combined anterior and posterior fusion

- It is used for thoracic, thoracolumbar, or lumbar curves with a poor prognosis.
- Because of the combined approach, the pseudoarthrosis rate is lower.
- Usually is performed under the same anesthetic.
- It is preferable to perform the anterior procedure first by removing an appropriate rib to expose the area of the curvature and then by removing the disk and growth plates. This provides greater mobility in the curvature, allowing a better correction.
- Posterior spine fusion is performed with or without instrumentation.
- Contraindicated in very young child with a kyphotic deformity.

Hemivertebra excision

- First performed in 1921 in Australia by Royle.
- Is indicated for patients younger than 5 years and who have development of a structural secondary curve with a fixed decompensation in which adequate alignment cannot be achieved with other procedures, like hemivertebra at the fourth or fifth lumbar level because no spine exists below the hemivertebrae to allow compensation.
- The hemivertebra is excised in two stages.
- The spine is approached both anteriorly and posteriorly.
- Depending on the age of the child and the size of the vertebrae, instrumentation is added.
- When it is not possible to add instrumentation, correction is maintained with a body cast with a leg extension, and the child usually is nonambulatory for 3-4 months.

Vertebrectomy

- It is the removal of two or more vertebrae in their entirety, including pedicles from both sides, laminae, and bodies.
- This is performed in order to create mobility but at the price of instability.
- This procedure is neurologically risky and must be accompanied with appropriate spinal cord monitoring and wake-up tests.
- It should be reserved for the most severe deformities and performed only by highly skilled spinal surgeons.

Congenital kyphosis

- Deformity in the sagittal plane resulting in an excessive flexion of the affected area.
- Due to failures of formation, segmentation or dislocation of the spine as a result of rotation.
- failures of formation may be classified by using three subclasses :

  1.Partial failure of formation of a vertebral element with a canal that is still properly aligned.

  2.Partial failure of formation with a malaligned canal.

  3.Complete failure offormation of a vertebral body.
- Defects of formation usually involve one level but can involve multiple levels.
- Most defects of formation occur in the thoracic or thoracolumbar segments of the spine.

Congenital kyphosis

- Failures of segmentation typically involve more than one level and usually present as an unsegmented bar.
- Rotatory dislocation of the spine, is an area of kyphosis found between two congenital scoliotic curves, both of which are lordotic and in opposite directions.
- Rotatory dislocation of the spine is usually found in the thoracic or thoracolumbar regions but can involve any vertebral segments.

Surgical treatment

- **Defects of formation**

  The main goal of treatment is prevention of paraplegia.

  .If patient is younger than 5 years, and less than 50° of kyphosis is present, posterior fusion alone will produce a desired outcome.

  .Postoperatively, the child is placed in a hyperextension cast and is kept nonambulatory for 3-4 months.

  .When the deformity is larger than 50° or when the child is older than 5 years, a combined anterior and posterior arthrodesis is mandatory.

  .The anterior fusion is performed first, with radical excision of the anterior longitudinal ligament, the disks, and the related ligaments. If possible, a distractor should be used to lengthen the anterior column, and structural bone graft from the rib or fibula should be placed anteriorly to maintain the achieved height.

- The anterior procedure is followed (either on the same day or a week after) by a posterior arthrodesis and compression instrumentation if bones are large enough to accept hooks and rods. Otherwise, the patient is managed with a hyperextension cast and bed rest for several months.

- **Defects of segmentation :**
- Defects of segmentation can be treated with a posterior fusion, which should include the entire kyphosis and one vertebra cephalad and caudad to the lesion.
- Because the deformity is rigid, instrumentation is not used

Congenital lordosis

- This defect is very uncommon and the least severe of the three abnormal curvatures.
- Defects of this kind create an abnormal extension of the spine.
- Three factors that may contribute to congenital lordosis :

  1.Posterior defect of segmentation with concomitant normal anterior development.

  2.Abnormal or lack of formation of the posterior elements.

  3.Compensatory deformity as a result of a kyphosis at a lower vertebral level.

- Most patients with a congenital lordosis exhibit some element of scoliosis (Winter et al).

Congenital lordosis management

- Treatment of congenital lordosis is purely operative.
- Two types of surgery are available, one for anterior fusion and the other for correction.
- The anterior fusion should include the entire involved area and 1 or 2 vertebrae cephalad and caudad to the lesion.
- Corrective operation is performed when the patient has a major deformity and, usually, loss of pulmonary function.
- A combined anterior and posterior approach is indicated in these cases.
- The goal is to improve the spinal alignment and pulmonary function.

CONGENITAL ANOMALIES OF THE CERVICAL SPINE

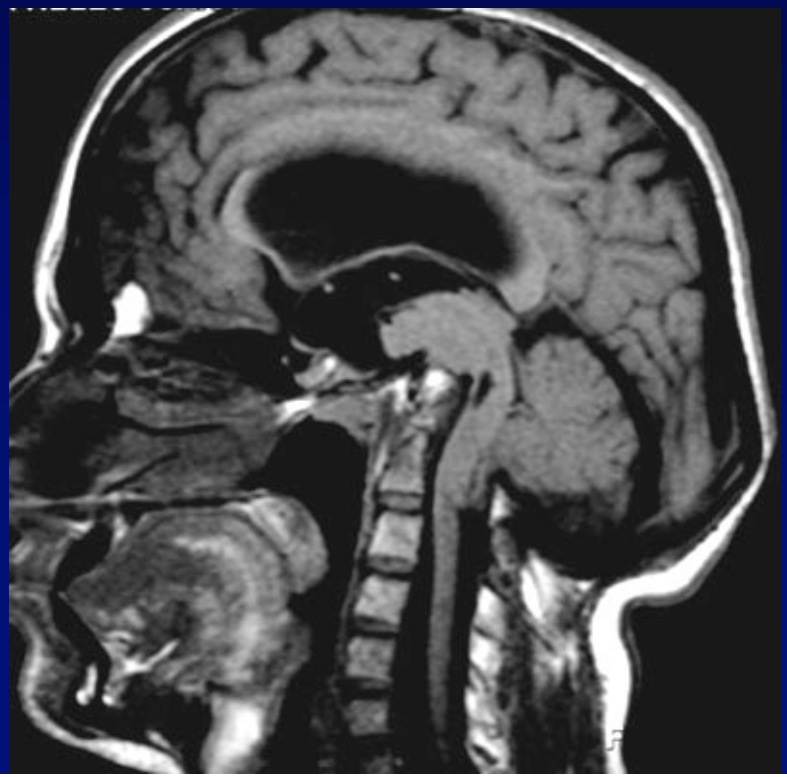
- **Basilar impression (invagination) :**
- The floor of the skull appears to be indented by the upper cervical spine; therefore, the tip of the odontoid is more cephalad.
- Two types of basilar impression exist:

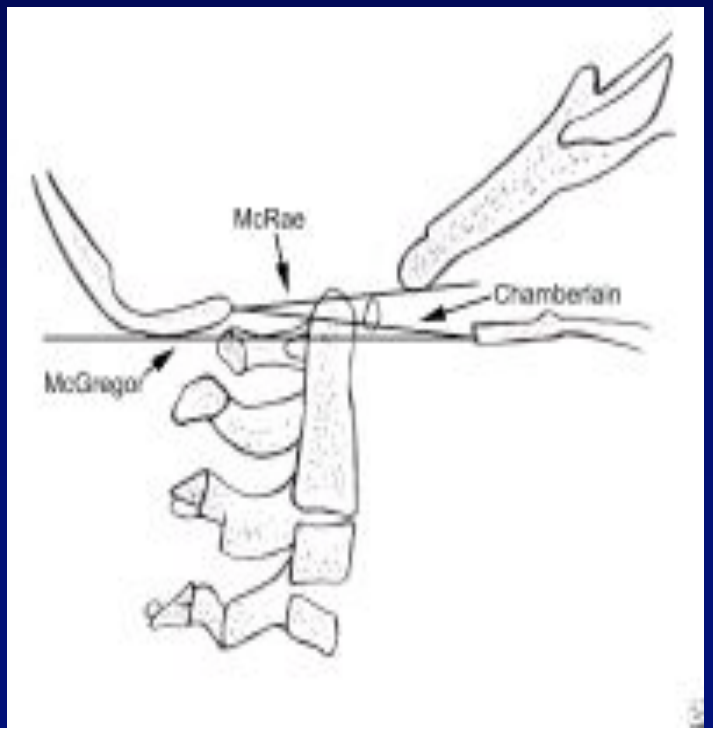
  (1) primary, a congenital abnormality often associated with other anomalies such as atlanto-occipital fusion, hypoplasia of the atlas, bifid posterior arch of atlas, odontoid abnormalities, Klippel-Feil syndrome, and Goldenhar syndrome.

  (2) secondary, a developmental condition usually attributed to softening of the bone, in which the deformity develops later in life.

. Motor and sensory disturbances are noted in 85% of individuals who are symptomatic

- Most affected patients remain asymptomatic until the second or third decade of life, when they may present with headache, neck ache, and neurologic compromise.
- McGregor's line is the best method for screening. Line is drawn from the upper surface of the posterior edge of the hard palate to the most caudad point of the occipital curve of the skull.
- The position of the tip of the odontoid is measured in relation to this base line and a distance of 4.5 mm above McGregor's line is considered to be on the extreme edge of reference ranges.





Treatment

- A hypermobile odontoid may require fusion in extension if the odontoid can be reduced. If the odontoid cannot be reduced, an anterior excision and stabilization in extension can be considered. Posterior approach may require suboccipital craniectomy and decompression of the posterior ring of C1 and possibly C2 with the release of tight dural bands. This is followed by fusion of the occiput to C2 or C3.

• Occipitocervical synostosis

- partial or complete congenital union between the atlas and the base of the occiput.
- It also is known as occipitalization of the atlas.
- Incidence ranges from 1.4-2.5 per 1000 children.
- M = F
- The following most common signs and symptoms occur in decreasing order of frequency:

  Pain in the occiput and neck
  Vertigo
  Unsteady gait
  Paresis of the limbs
  Paresthesias
  Speech disturbances
  Hoarseness
  Double vision
  Syncope
  Interference with swallowing

- Other clinical findings include a short broad neck, a low hairline, torticollis, a high scapula, and restricted neck movements.
- Other associated anomalies occasionally seen include dwarfism, funnel chest, pes cavus, syndactylies, jaw anomalies, cleft palate, congenital ear deformities, hypospadias, and, sometimes, genitourinary tract defects.
- CT scan, and MRI may be necessary to clarify the pathologic condition.

- **Odontoid anomalies :**

- Range from complete absence (aplasia) to partial absence (hypoplasia) to separate odontoid process, or os odontoideum.
- May lead to atlantoaxial instability and may cause neurologic deficit and even death.
- Partial or complete absence of the dens may be either congenital or acquired. This extremely rare anomaly may be recognized from birth onward and is best seen in the open-mouth view.
- In os odontoideum, there is a jointlike articulation between the odontoid and the body of the axis, which appears radiologically as a wide radiolucent gap. This gap may be confused with a normal finding in patients younger than 5 years.
- Surgical stabilization is indicated if neurologic involvement is present with more than 10 mm of instability on supervised flexion-extension films.
- Posterior cervical fusion of C1-C2, with wire fixation and an iliac bone graft.

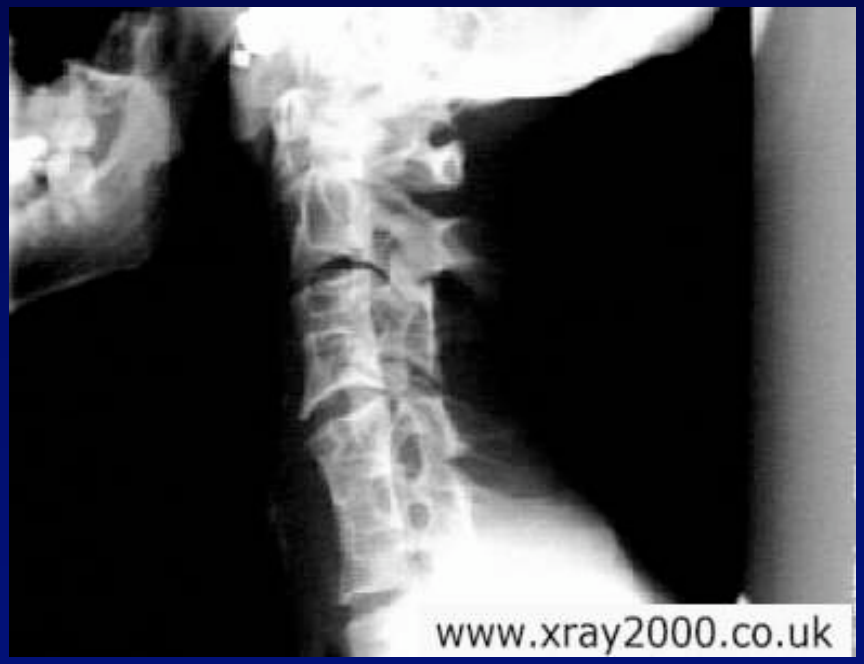
Klippel-Feil syndrome

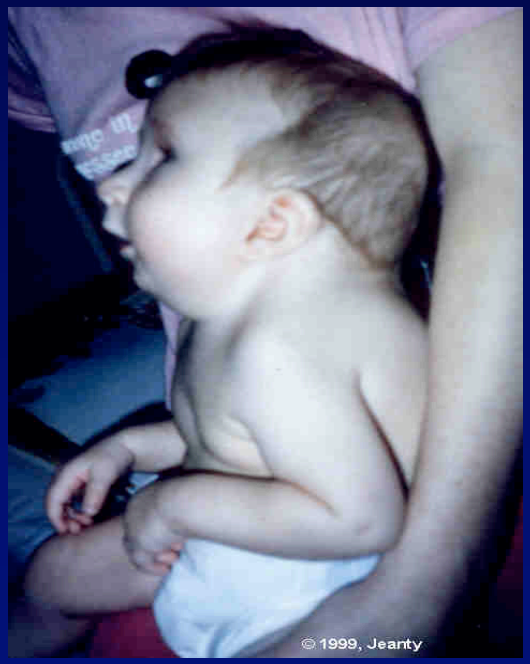
- Involves the congenital fusion of two or more cervical vertebrae.
- F>M
- 1 in 42,000 births
- The abnormality is the result of a failure of proper segmentation of vertebrae
- Patients with Klippel-Feil syndrome have a shortened neck with a low posterior neckline and a diminished ability to move in the affected area.

- Klippel-Feil syndrome is classified into three categories:
- **Type I** comprises fusion of many cervical and upper thoracic vertebrae into bony blocks.
- **Type II** has fusion at only one or two vertebrae and may associate hemivertebrae and fusion of the occipito-atlantoic joint.
- **Type III** has both cervical fusion and lower thoracic or lumbar fusion.

Klippel-Feil syndrome

- 60% of patients with this syndrome have scoliosis, and 35% have an abnormality in the urinary system.
- 30% of patients present with impaired hearing.





Congenital spondylolisthesis

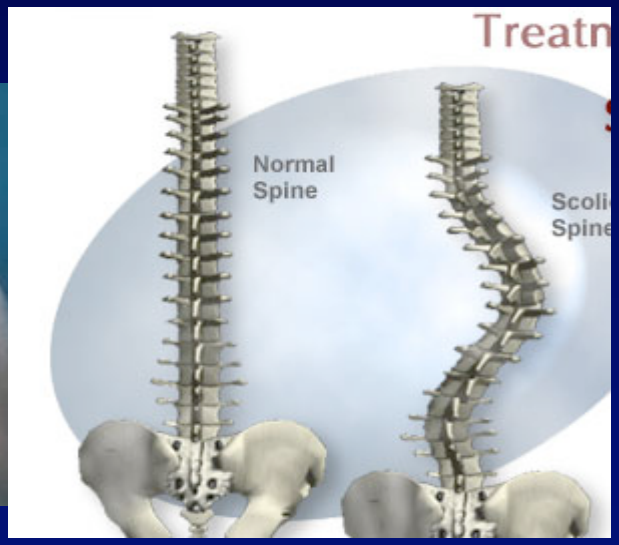
- Condition that involves a forward slip of a vertebra or vertebrae in relation to the rest of the spinal column.
- Typically, this defect occurs between the L5 vertebra and S1. However, spondylolisthesis has also been seen between L4 and L5 vertebral levels and may even occur in the cervical region.

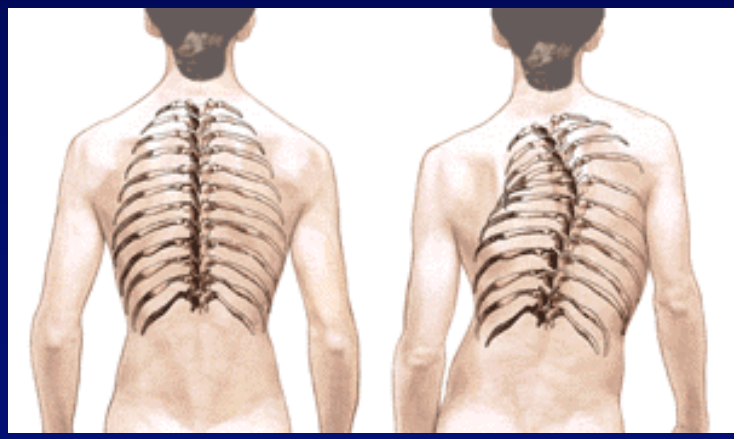
SYNDROMES ASSOCIATED WITH CONGENITAL SPINAL DEFORMITY :

- Down syndrome (trisomy 21 syndrome)
- Deletion 5p syndrome (chromosomal number 5 syndrome)
- Kabuki syndrome.
- Noonan syndrome (Turnerlike syndrome).
- Aarskog syndrome.
- Cervico-oculo-acoustic syndrome (Wildervanck syndrome).
- MURCS association (müllerian duct, renal, and cervical vertebral defects).
- VACTERL association.
- Jarcho-Levin syndrome (spondylothoracic dysplasia).
- Proteus syndrome.

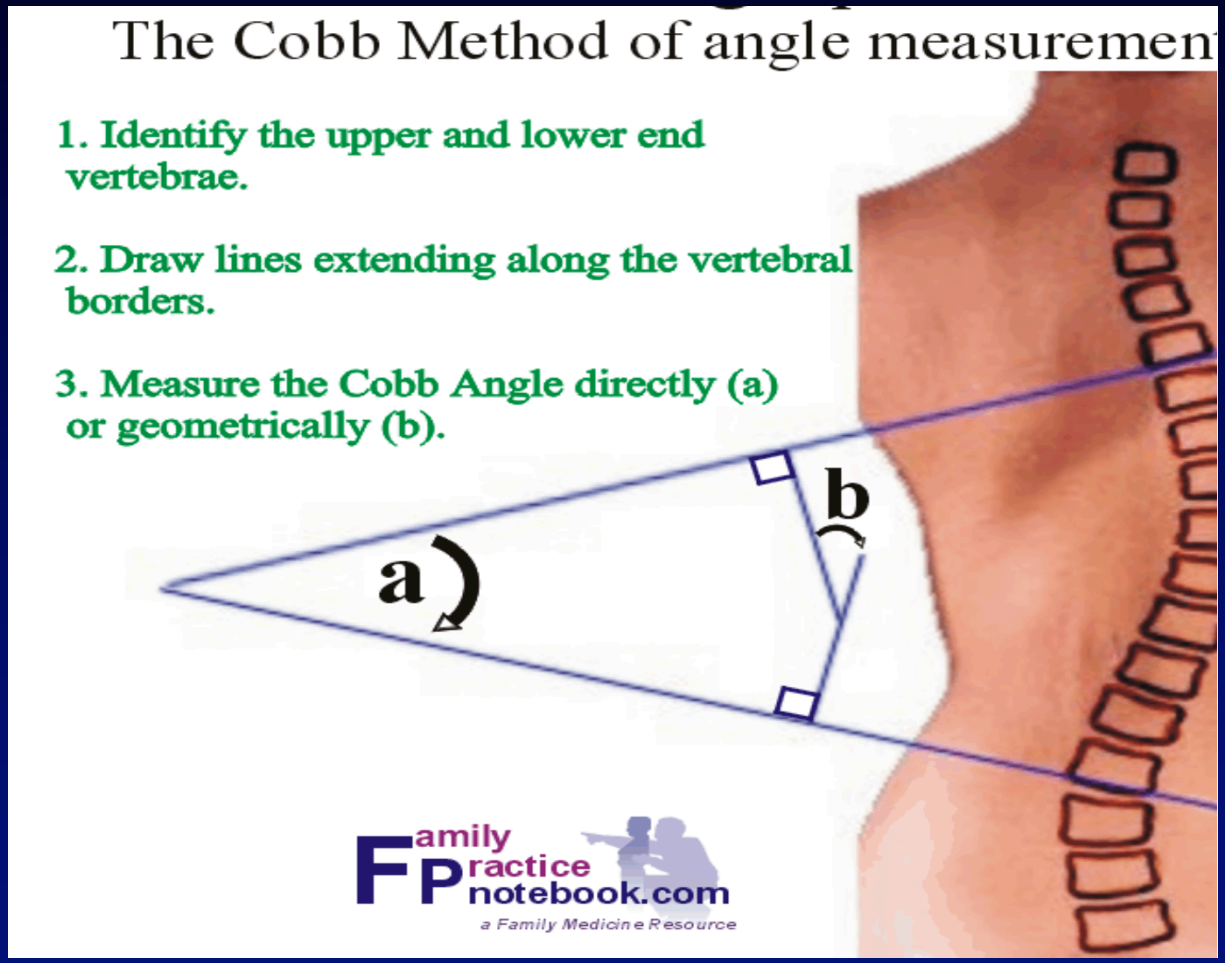
Scoliosis

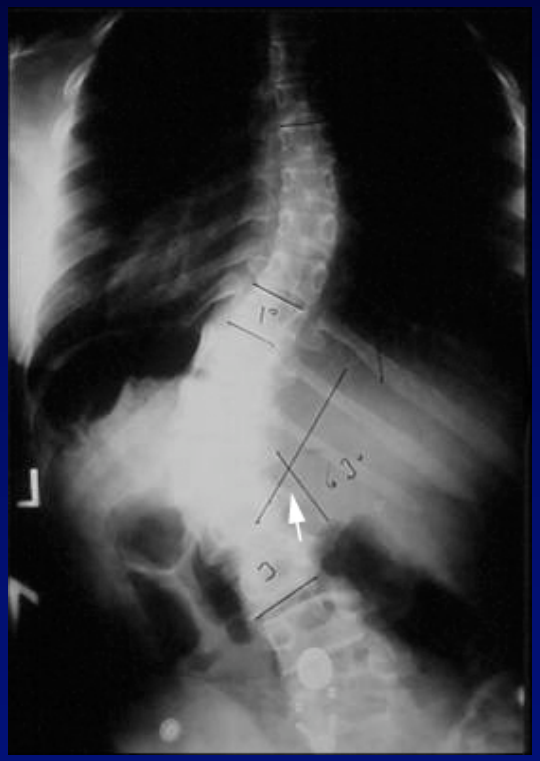
- Scoliosis is not a diagnosis, but a description of a structural alteration that occurs in a variety of conditions. Progression of the curvature during periods of rapid growth may result in significant deformity, which may be accompanied by cardiopulmonary compromise.
- Scoliosis is defined as curvature of the spine in the coronal plane.
- 10º of curvature (as measured by the Cobb angle) defines a scoliosis.

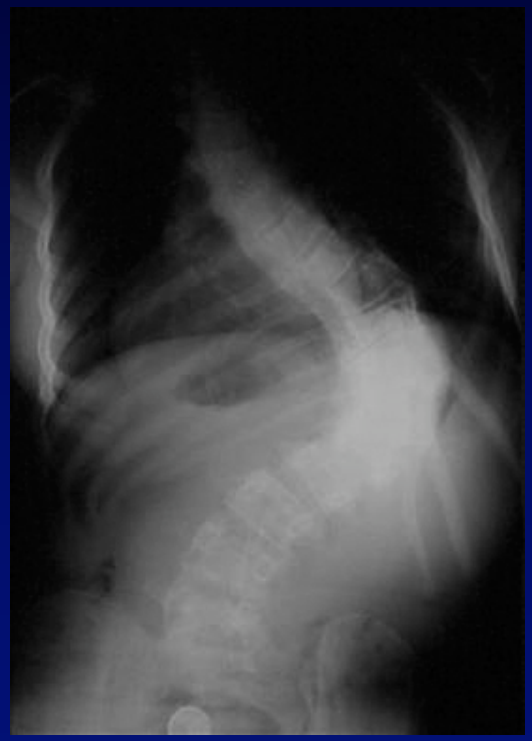




- **Cobb angle**
- The degree of curvature in the coronal plane is measured according to the method of Cobb. The Cobb angle is formed by the intersection of a line parallel to the superior end plate of the most cephalad vertebra in a particular curve, with the line parallel to the inferior endplate of the most caudad vertebra of the curve.
- From a practical point of view, when these two lines are drawn, their intersection often is outside the border of the actual film. Thus, by convention, perpendiculars to the parallels are drawn, and the angle between their intersection is measured.


The Cobb Method of angle measuremen
1. Identify the upper and lower end vertebrae.
2. Draw lines extending along the vertebral borders.
3. Measure the Cobb Angle directly (a) or geometrically (b).
a)
b
Family Practice notebook.com
a Family Medicine Resource





Etiologic classification of scoliosis

- **Idiopathic***
  Early-onsetInfantile
  Juvenile
  Late-onset (Adolescent)

.Congenital vertebral defect* Hemivertebrae
  Wedge vertebrae
  Failure of segmentation
  Unilateral (bar)
  Bilateral (fusion)
  Fused ribs

- **Neuromuscular***
- **<u>Neuropathy</u>**
  Cerebral palsy
  Myelomeningocele
  Tethered spinal cord
  Spinal cord injury
  Syringomyelia
  Diastematomyelia
  Friedreich ataxia
  Charcot-Marie Tooth disease
  Juvenile spinal muscle atrophy
  Poliomyelitis

- **Myopathy**

  Duchenne muscular dystrophy
  Nemaline Myopathy
  Facioscapulohumeral dystrophy
  Limb-girdle muscular dystrophy
  Arthrogryposis

- **Trauma**

  Direct vertebral trauma
  Irradiation
  Extravertebral trauma

. **Mesenchymal origin**

  Marfan syndrome
  Ehlers-Danlos syndrome
  Congenital laxity of the joints
  Homocystinuria

- **Tumors**

Vertebral
  Ostoid osteoma
  Osteoblastoma
Intraspinal
Extramedullary (eg, neurofibroma)
Intramedullary (eg, astrocytoma)

- **Miscellaneous**

Vertebral body infection
Rickets
Osteogenesis imperfecta
Schueurmann disease
Achondroplasia
Klippel-Feil syndrome
Sprengel deformity
Cleidocranial dysostosis
Hyperphosphatasia
Hypervitaminosis A

Hypothyroidism
Dysautonomia
Juvenile rheumatoid arthritis
Mucopolysaccharidoses

CLINICAL EVALUATION

- Evaluation of the adolescent with scoliosis has several objectives, including identification of an underlying etiology (ie, excluding nonidiopathic causes, assessment of the magnitude of the curve and need for radiographs, and determining the risk of progression, which influences management decisions.
- **History**
- When was the deformity first noted and who noted it?
- What is the rate of progression? Is there associated pain?
- The presence of significant pain (pain that limits activities or requires frequent analgesia) increases the likelihood of nonidiopathic etiology.
- Are there associated symptoms suggestive of a neuromuscular etiology (eg, muscle weakness, bowel or bladder problems, headache, neck pain). Neurologic symptoms increase the likelihood of nonidiopathic causes and require additional evaluation.

History cont.

- Does the patient have shortness of breath or difficulty breathing? Severe thoracic scoliosis may affect pulmonary function.
- What is the patient's growth trajectory and has the pubertal growth spurt begun? This information helps to estimate remaining linear growth and need for intervention.
- Has the patient entered puberty?
- For female patients, has menarche occurred? If so, when? Girls continue to grow at decreased velocity for approximately one year after menarche and complete linear growth by 24 months after menarche.
- Is there a history of lower limb fracture, joint infection, or arthritis (which may result in leg length discrepancy)?
- Is there a family history of scoliosis?

- **General examination**
- Measurement of the patient's height
- Excessive skin or joint laxity.
- Asymmetry on bilateral palpation of the iliac crests and posterior inferior iliac spines with the patient in the standing position with the hips and knees fully extended suggests leg length discrepancy.
- Examination of the skin for café-au-lait spots and axillary freckling (suggestive of neurofibromatosis)

  vascular, hypopigmented, or hypopigmented lesions or a patch of hair overlying the spine (which may be associated with spinal dysraphism)

  and dimpling in the lumbosacral area (which may be associated with intraspinal tumor).

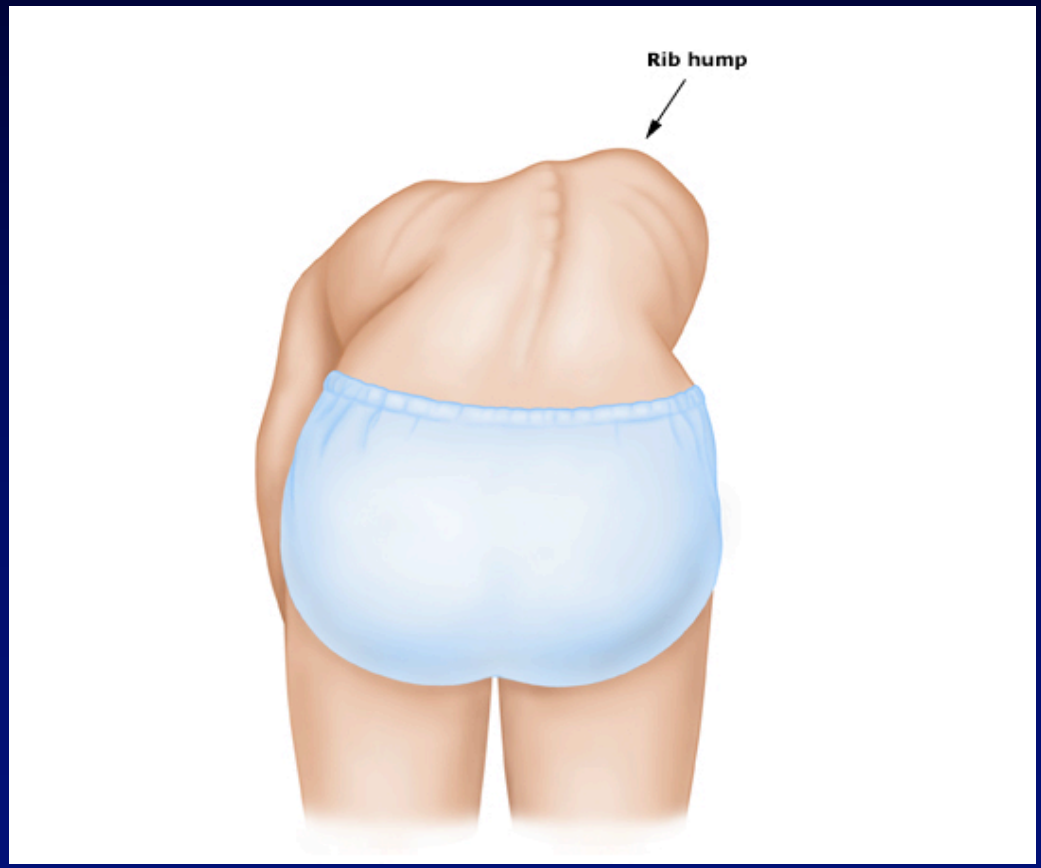
- The feet should be examined for high arches (pes cavus) and hammer or claw toes, which are suggestive of neuromuscular disease.
- A full neurologic examination, including examination of the reflexes should be performed. The abdominal reflex is especially important, since an absent abdominal reflex is sometimes indicative of subtle intraspinal pathology.
- **Scoliosis examination**
- **Inspection**
- Viewing a standing patient from behind

  Differences in the height of the shoulders or scapulae, waistline symmetry, Distance arms hang from the trunk may be appreciated on closer inspection The patient's head should appear "balanced" over the center of the sacrum and not shifted to one side or the other.

- Viewed from the side

  The patient should have the normal profile of thoracic kyphosis and lumbar lordosis; decreased thoracic kyphosis may be seen.

- **Forward bend test** The Adams forward bend test is performed by observing the patient from the back while he or she bends forward at the waist until the spine becomes parallel to the horizontal plane, with feet together, knees straight ahead, and arms hanging free. In a patient with scoliosis, a thoracic (rib) or lumbar (loin) prominence on one side will be evident


Rib hump

- # Use of a scoliometer

- A scoliometer is a device used for scoliosis screening and quantification of trunk rotation. Scoliometer measurements can help in determining which patients need radiographs, but should not be used in the absence of radiographs to make decisions regarding bracing or surgery.
- A scoliometer is basically a version of a carpenter's level that measures the angle of trunk rotation. The scoliometer is run along the patient's spine from cephalad to caudad while the patient is in the position assumed for the Adams forward bend test. If a rotational prominence (ie, rib or loin hump) is present, the ball in the scoliometer deviates from the center of the device. If there is a right rib prominence, for example, the right side of the scoliometer tips upward, and the ball deviates to the left.


Scoliometer


Scoliometer

RADIOGRAPHIC EVALUATION

- **Indications**

1. Scoliometer reading of 7º, since curves of this magnitude may require treatment.
2. Clinically evident scoliosis on physical examination.
3. Asymmetry on physical examination in skeletally immature children or in combination with a family history of scoliosis.
4. Monitoring progression in patients with previously diagnosed with scoliosis.

5. Magnetic resonance (MR) imaging may be indicated in patients with scoliosis and clinical or plain radiographic findings suggestive of intraspinal pathology (tumor, dysraphism, infection). These findings include:

- Associated neurologic signs or symptoms, headache, neck pain, absence of abdominal reflex, and midline skin lesions (vascular, pigmentary, hair patch).
- Complaints of significant associated pain.
- Early age of onset (before 10 years) with rapid progression.
- Left-sided thoracic curves in children with abnormal neurologic examination.
- Abnormalities on plain radiographs that suggest congenital scoliosis or intraspinal pathology (eg, widening of the interpediculate space or erosion of the pedicles) or increased kyphosis.

Radiographic findings :

1. General

- Soft tissue abnormalities (eg, paraspinal mass)
- Wedged vertebrae or hemivertebrae (indicative of congenital scoliosis)
- Vertebral body lucency (suggestive of bone tumor)
- Widening of the interpediculate space or erosion of the pedicles (suggestive of a spinal cord tumor, syringomyelia, diastematomyelia, or spinal dysraphism)

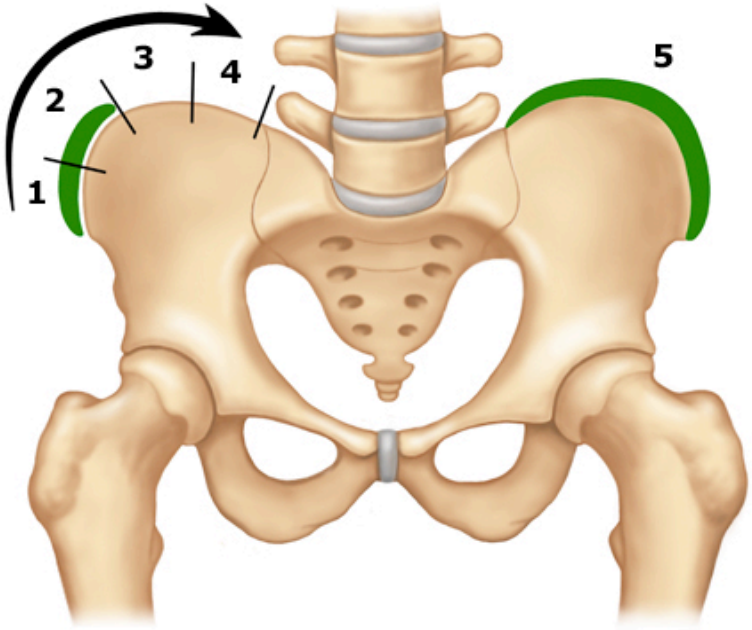
2. Curve pattern

- The direction of the curve (right or left) is defined by its convexity.
- The location is defined by the vertebra that is most deviated and rotated from midline (the apical vertebra).

3. Cobb angle

4. Risser sign

- grading of the degree to which the iliac apophysis has undergone ossification and fusion, is used to assess skeletal maturity.
- The iliac apophysis ossifies in a stepwise fashion from anterolateral to posteromedia



COMPLICATIONS OF SURGERY :

- Bleeding, coagulopathy and DIC.
- Spinal cord injury.
- Postoperative activity level and the possible hazards of trauma.
- Infections.
- Crankshaft phenomenon : It may occur following posterior spinal fusion of idiopathic scoliosis in patients who have significant anterior spinal growth remaining (highest in patients younger than 10 years).
- Future low back pain, related to the inferior (caudad) extent of spinal fusion.
- Complications related to surgical approaches to scoliosis : chylothorax and tension pneumothorax.
- Pseudarthrosis.

Differences between the Paediatric and Adult spine :

- **In general**
- the vertebral ossification centers are incompletely ossified early in childhood.
- Disks are thicker and have a higher water content than those in adult.
- The spinal canal and neural foramina are larger.
- The overall signal intensity of v. bodies is lower than that of adult spine on T1-weighted image due to abundance of red marrow.
- **Full-term Infant**
- The overall size of v. body is small relative to spinal canal.
- Spinal cord ends @ L2 level.
- No Lumbar Lordosis.

- **Age 3 months**
- The osseous component of the v. body increased and the amount of hyaline cartilage decreased, giving the v. body rectangular shape.
- The ossification centres begin to gain in signal intensity starting at the end plates and progressing centrally.
- **Age 2 years**
- Spine start show normal sagittal alignment, most likely due to wt. bearing.
- The ossified portion of v. body increases and begins to assume adult appearance, with near complete ossification of pedicles and articular p.
- The disk space and nucleus p. become longer and thinner.
- C. end plate decreased in size.
- Neural foramen begins to take adult appearance.

- **Age 10 years**
- Sagittal alignment resembles that of an adult.
- Ossification of the v. bodies and post. Elements is nearly complete.
- Decrease in spinal canal diameter.
- The v. bodies develop concave superior and inferior contours.
- The nucleus p. become smaller.
- The neural foramina continue to narrow inferiorly.

Idiopathic Scoliosis:

- Scoliosis is commonly defined as greater than 10° of lateral deviation of the spine from its central axis.
- It is grossly oversimplified as mere lateral deviation of the spine, when in reality, it is a complex 3-dimensional deformity.
- J.I.P. James is credited with classifying idiopathic scoliosis according to the age of the patient at the time of diagnosis (James, 1954).
- Using his classification system,

1. infantile idiopathic scoliosis when children are younger than 3 years.
2. juvenile idiopathic scoliosis when children are aged 3-10 years.
3. adolescent idiopathic scoliosis children are older than 10 years.

- **Frequency:**
- The prevalence of idiopathic scoliosis (Cobb angle >10°) is 0.5% (76 of 15,799 patients) (Stirling, 1996).

- Other studies using the 10° definition of scoliosis have placed the overall prevalence in the 1.9-3.0% range (Albanese, 2002).

- Most patients with idiopathic scoliosis are female.

- **Etiology:** The precise etiology of idiopathic scoliosis remains unknown.
- A primary muscle disorder .
- An elastic fiber system defect (abnormal fibrillin metabolism).
- Disorganized skeletal growth.
- Genetic. More than 90% of monozygotic twins and more than 60% of dizygotic twins demonstrate concordance regarding their idiopathic scoliosis (Inoue, 1998).

  Some evidence has also directed attention to portions of chromosomes 6, 10, and 18 as possible scoliosis-related loci (Wise, 2000).

- **Clinical presentations:**
- Deformity: This may be patient or family perception of asymmetry about the shoulders, waist, or rib cage.
- Back pain 23%.
- Shoulder unleveling.
- protruding scapulae.
- Physical examination should include assessment of lower and upper extremity reflexes. Abdominal reflex patterns should also be assessed. The presence or absence of hamstring tightness should be investigated, and screening should be performed for ataxia and/or poor balance.

- Treatment :

The main treatment options for idiopathic scoliosis may be summarized as "the 3 O's":

(1) Observation
(2) Orthosis
(3) Operative intervention.

Infantile idiopathic scoliosis :

- <3 y at the time of diagnosis.
- It is the only type of idiopathic scoliosis whose most common curve pattern is left thoracic.
- It is the only type of scoliosis that is more common in boys.
- It is also the only type of idiopathic scoliosis shows significant spontaneous resolution. Reported spontaneous resolution rates range from 20-92% (James, 1954; Lloyd-Roberts, 1965).
- It is more common in European patients.
- Management outline for infantile idiopathic scoliosis may be as follows:
- Curves less than 25° with an RVAD less than 20° are observed and monitored with spinal radiographs at regular intervals.
- Curves exceeding these parameters are typically braced, with some consideration given to the value of intermittent casting.
- Surgery is considered for curves not adequately controlled with nonoperative measures.

- Prediction of curve progression in infantile idiopathic scoliosis has been tied to assessment of the rib vertebral angle difference (RVAD) originally described by Mehta in 1972 (Mehta, 1972). As described by Mehta, this measurement is carried out at the apical vertebra of the curve.
- The rib-vertebra angle difference is the difference between the rib-vertebral angle on the convexity of the curve subtracted from that on the concavity and may be either a positive or negative value.

Juvenile idiopathic scoliosis :

- It is more common in females.
- Its most common curve pattern is a right thoracic curve.
- It is considered to be a malignant subtype of adolescent idiopathic scoliosis.

- One potential treatment algorithm for juvenile idiopathic scoliosis is as follows:
- Observation for curves less than 25° with follow-up radiographs at regular intervals.
- Bracing for curves that range from 25-40° and at least consideration of bracing (based on curve flexibility) for curves from 40-50°
- Bracing for smaller curves that demonstrate rapid progression to the 20-25° range.
- Surgical intervention for inflexible curves that exceed 40° or virtually any curve that exceeds 50°.

Adolescent idiopathic scoliosis :

- It is the most common type of idiopathic scoliosis and the most common type of scoliosis overall.
- Progressive curvature may be predicted by a combination of physiologic and skeletal maturity factors and curve magnitude, i.e. small curves in more mature patients have a substantially lower risk of progression (about 2%) than larger curves in more immature patients, in whom the risk is much higher (risk may approach or exceed 70%).
- Treatment recommendations for adolescent idiopathic scoliosis are driven almost totally by curve magnitude. Observation for curves less than 30°, bracing of curves that reach the 30-40° range, and consideration of surgery for curves that exceed 40°.
- This amounts to a 10° window between observation and major spinal surgery. (10° is a commonly discussed margin of error for measuring such scoliotic curves). ??

- When it comes to surgical considerations, patients with adolescent idiopathic scoliosis may be functionally subdivided into those patients in whom significant anterior spinal growth is a concern and those in whom it is not. This amounts to a quantification of risk of development of the complication known as crankshaft phenomenon. This can have a major impact on the surgical treatment plan in that a child at significant risk for crankshaft phenomenon will require an anterior spinal fusion procedure.