

# SUMMARY

Spinal dysraphism is a group of congenital anomalies of the spine in which the midline structures fail to fuse.

Spinal dysraphism includes; two groups spina bifida aperta or open neural tube defects, and spina bifida occulta or closed neural tube defects which result in tethered cord syndrome and commonly manifested in the skin by hypertrichosis, hemangioma, pigmentation, atrophic skin and subcutaneous lipoma.

Embryologically the abnormality manifests between the third and fourth week of gestation. At this point in development the neural plate folds into neural tube a process called neurulation.

Different aetiological factors are responsible for the spina bifida which includes. Genetic factors most commonly through multifactorial inheritance, prevention of genetic factors through genetic counselling and proper prenatal diagnosis which include (1) Antenatal ultrasound (2) maternal serum and alpha fetoprotein.

Other factors or non genetic factors nutritional factors especially deficiency of folic acid during pregnancy also other numerous teratogens as valproic acid, alcohol and viral infections.

So through this aetiological factors we send a message for the obstetricians and Gynaecologists for proper antenatal care and supplementation of maternal diet.

The pathogenesis of spina bifida aperta is discussed according to different theories which includes developmental arrest, over growth hypothesis, hydrodynamic theory, neuroschisis theory and centre migration theory.

The pathogenesis of tethered spinal cord syndrome is due to one factor or combination of many factors such as traction, compression and rarely inflammation. The underlying mechanism is due to impairment of oxidative metabolism of the spinal cord. Neurological dysfunction may result from the combination of stretching and repetitive trauma to the fixed cord that may occur during the process of normal flexion and extension of the spine in day to day activity.

The different pathological types of spina bifida aperta includes, myelomeningocele, meningocele, hemimyelomeningocele, myelocystocele.

The different pathological varieties of occult spinal dysraphism includes spinal cord lipoma, lipomyelomeningocele, split cord malformation, congenital dermal sinus, abnormal film terminale, lateral and anterior spinal meningocele and neurenteric cysts.

Clinical diagnosis of spina bifida aperta through examination of newly born assessment of motor and sensory functions. Evaluation of sphincteric action and deformities of lower limbs and vertebral column.

Hydrocephalus and Arnold Chiari malformation are common features of SBA occurring in about 80 to 90% of cases.

In tethered cord syndrome the patients presented by sphincteric dysfunctions, motor or sensory disorders in the legs, pain, cutaneous manifestations or musculoskeletal abnormalities.

Radiological investigations of spinal dysraphism are very important. The methods of investigations are plain X-ray spine that may show spina bifidia or other skeletal abnormalities, myelography or computerized tomography that detect the level of the conus and the cause of tethering. Magnetic resonance, imaging is the investigation of choice if available, the site and extent of tethering and the presence of other anomalies can be identified.

Early surgery of spina bifida aperta will preserve all neural tissue with special treatment but hydrocephalus and arnold chiari malformation surgery requires proper neonatal anaesthesia and proper positioning and handling of cases of spina bifida.

Surgery of tethered cord syndrome should be done as early as possible to prevent further neurological deficit that may result from acute tethering of the cord due to hyperflexion or hyper-extension of the spine. The difficulty of the operation for short thick filum terminale is much easier than the presence of adhesion bands or other anomalies. The aim of surgery is to release the cord from the tethering cause before any damage to the neural element.