I_❤_NEUROSURGERY INITIATIVE

SPINAL AND CRANIAL DYSRAPHISM

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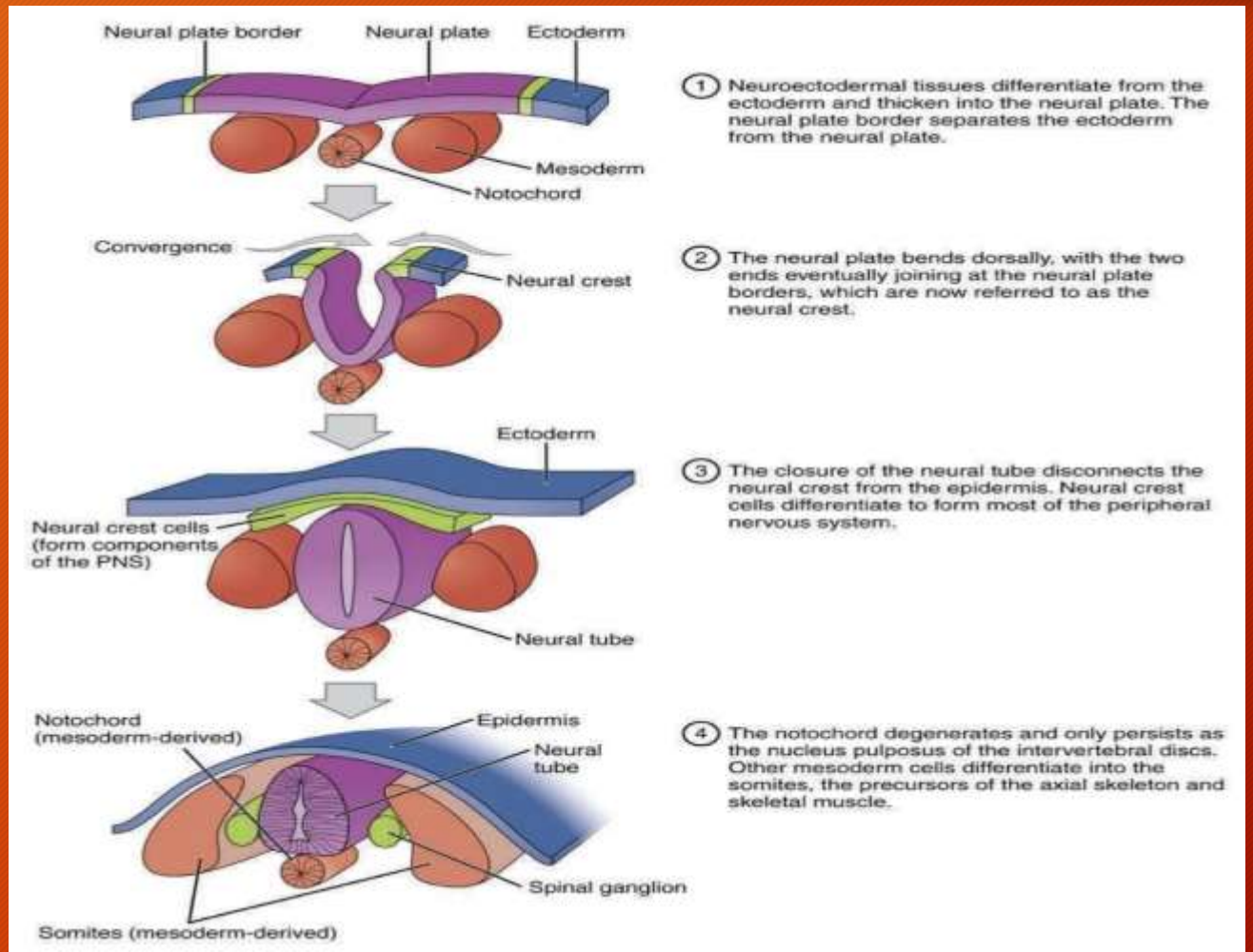
EMBRIOLOGY

- **During** development of the embryo, the neural tube forms as a fold from ectoderm, then gets separated from the uniting ectodermal layer by the growth of mesoderm from each side. The neural tube will form the nervous tissue, the mesoderm will form basically the coverings including dura and bone, and the ectoderm will form the skin.

EMBERIOLOGY
THE NEURAL TUBE



WHAT IS DYSRAPHISM?

- Any failure for this tube to form, or for the mesoderm and ectoderm to meet at all will result in dysraphism which literally means “defective fusion”, the word “rhaphe” comes from Greek language and means suture.

DYSRAPHISM

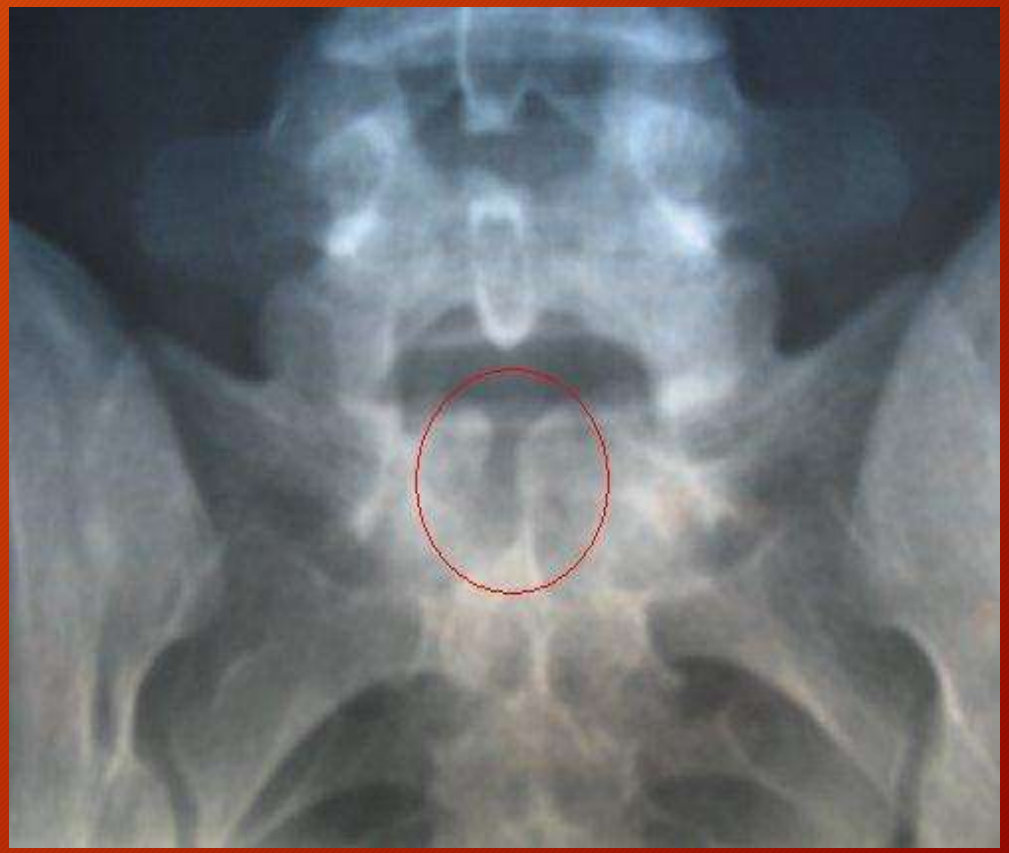
- The degree of failure of union in these layers will determine the severity of the resulting congenital defect from the mildest form of skin dimple in the midline of the lower back or the absence of a single spinous process, to the severest form of rachischisis where there is complete failure of the neural tube to close and the CSF of the central canal pours on the skin.

DYSRAPHISM

- These congenital abnormalities will be found in the midline of the body along its “raphe”, so we encounter them mainly in the back and at the nape of the neck in the occipital region, but we can find some in the roof of the pharynx or at the bregma

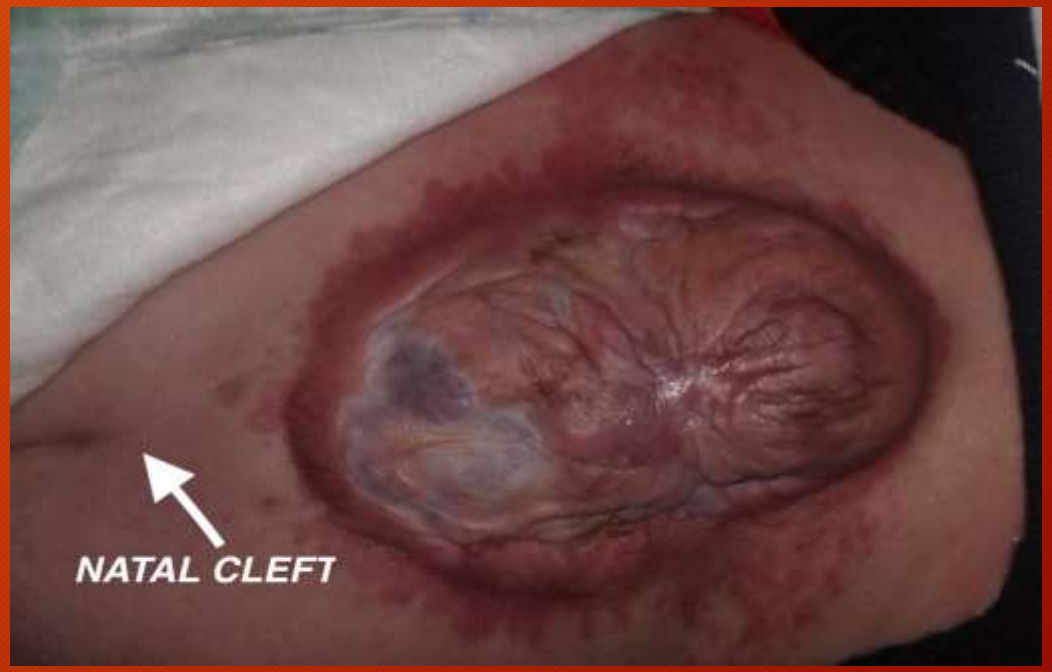
TYPES

- ***Spina bifida occulta***, in which there is no apparent defect on the surface of the body, but one may detect defects in the spinous process or laminae on imaging . The person is usually unaware of these and has no complaints. These anomalies will be discovered accidentally while the person is being imaged for some other problem.



TYPES

- ***Spina bifida aperta***, in which we can detect an anomaly on the surface of the skin. We will discuss two of these namely, meningocele and myelomeningocele



CRANIAL DYSRAPHISM ENCEPHALOCELE

- The same type of spinal defect can occur in relation to the skull and brain leading to encephaloceles. Some of these appear at the “bregma” and some in the nasopharynx, resembling polyps. Trying to “excise” them or aspirate them by non-neurosurgeons lead to disastrous results.

RISK FACTORS

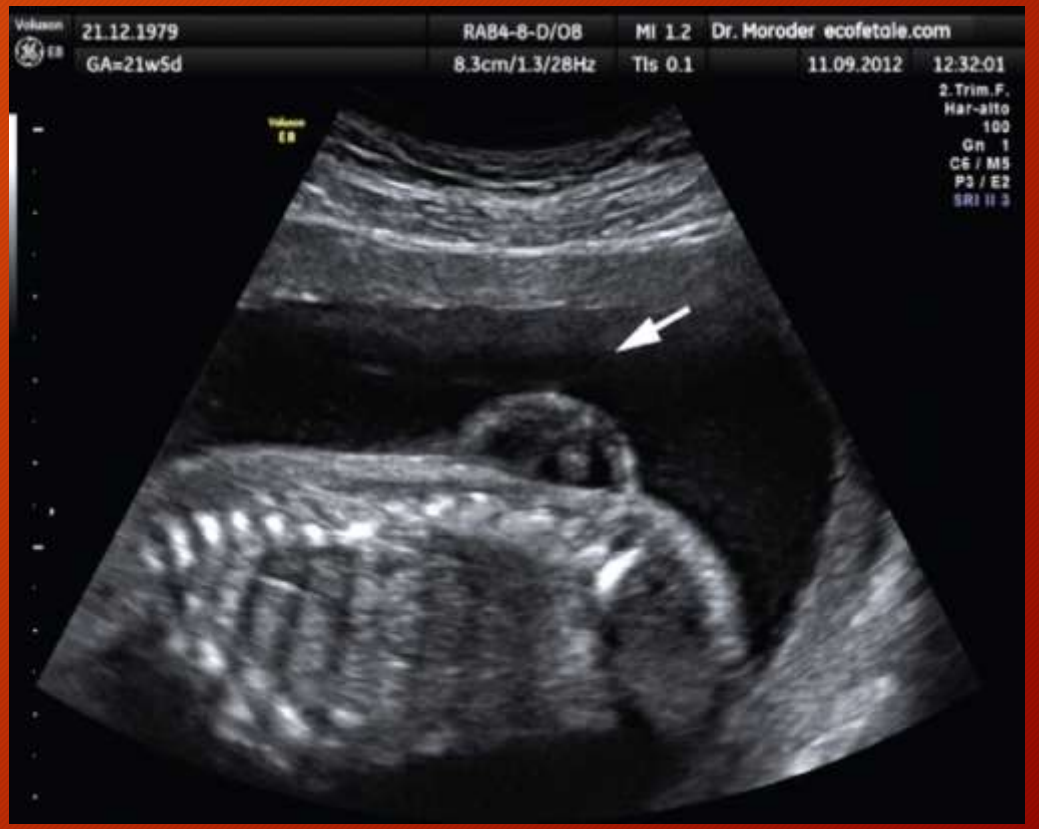
- There are many factors which influence the development of such anomalies (of the second category), including familial genetic causes, nutritional causes like folate deficiencies, and the intake of certain medication especially antiepileptic drugs. The risk increases with marrying into close family especially cousins.

INCIDENCE

- The incidence varies between countries but usually ranges between 1-2/1000 live births. This incidence double 10-fold if the family has one previous child with the anomaly, and 50-fold if it has two afflicted members. The embryogenic aberration occurs during the 4^{th} week of gestation, the same time during which the kidneys, heart and bones are formed. This is why we usually find associations between defects in these four systems

PRENATAL DIAGNOSIS

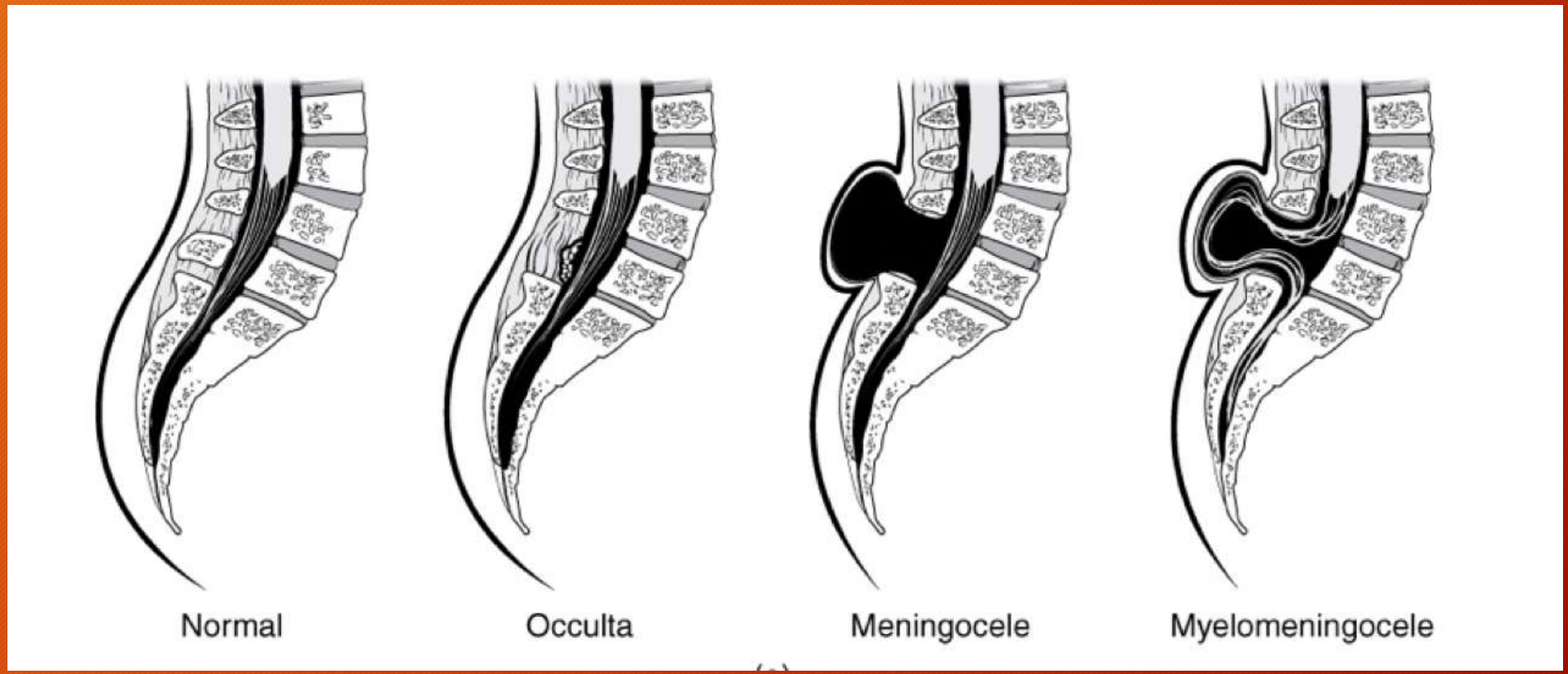
- Meningoceles and myelomeningoceles as well as encephaloceles could be detected before birth on routine ultrasonography and become a cause of concern regarding management. On many occasions encephaloceles were the cause of obstructed labor, the head and the large cyst being unable to be delivered normally.



THE ASSOCIATTION OF SPINA BIFIDA AND HYDROCEPALUS

- Myelomeningoceles may be associated with hydrocephalus in as much as 10% at birth, but around 80% will develop hydrocephalus in the first year of life. These patients usually have an associated Arnold-Chiari type 2 malformation, and many have an associated congenital aqueduct stenosis

TYPES



TYPES

MENINGOCELE

- In this condition, there is no involvement of the actual nervous tissue. The dura filled with CSF protrudes through a defect in the posterior arch of a vertebra, usually in the form of a narrow neck, to produce a cystic swelling in the midline of the back (usually lumbar or lumbosacral but could be dorsolumbar). The skin covering the cyst could be normal. The cyst will not contain any nervous tissue

TYPES

MYELOMENINGOCELE

- Here in addition to what was described for the meningocele, there is an added element of nervous elements herniating into the sac or being attached or adherent to its walls. Usually this nervous tissue is nonfunctioning or non-viable, leading to neurological deficits corresponding to the involved nervous element, which could be cord and roots if the myelomeningocele is dorsolumbar, or just roots if it were lumbosacral.

MANAGEMENT

- If born in the hospital they are already admitted, but if seen in the emergency room they should be admitted and managed according to their maturity.
- With regard to the neural defect it should be dealt with within the first 24-48 hours and sooner if it had already ruptured for fear of infection leading to meningitis. The baby is nursed on his abdomen and the defect covered with gauze soaked in saline. If it had already ruptured antibiotics should be administered.

MANAGEMENT

- Examination should determine the **lowest neurological function detected.** The power of the lower limbs determined, reflexes tested and the anus examined for tone. The head circumference should be measured and the anterior fontanelle examined for fullness. Ultrasound could be used to detect ventricular size.
- If the baby is premature assessment of his pulmonary function should be performed. It must be remembered that these patients may have poly cystic kidney disease or cardiac anomalies.

MANAGEMENT

- The aim of the neurosurgeon is to close the defect. The surgery is called repair of meningocele or myelomeningocele. It is easier to repair a meningoceles. The neck of the sac is excised, the dura is repaired, and a layer is fashioned from the lumbar fascia to cover the defect site and then, the excess skin excised.
- In myelomeningoceles, the task of repair is more difficult. The dura has to be freed and refashioned, the nervous elements have to be dissected free if possible, fascia from the adjacent lumbar area could be used to support the dura after closure.

MANAGEMENT

- Recent advances in fetal surgery, allowed trained neurosurgeons to manage these defects intrauterine. Avoiding the harmful effects of the amniotic fluid on the exposed neural tissue (if present), and also avoiding contamination and rupture during delivery.
- Due to the high percentage of associated hydrocephalus, and in cases in which the hydrocephalus is apparent, surgeons tend to shunt the ventricle in the same sitting. This will help in lowering the CSF pressure in the closed sac and reduce the chance of post-operative CSF leak. In those patients with no apparent hydrocephalus, the shunting process could be delayed for few days