Meningoencephalocele

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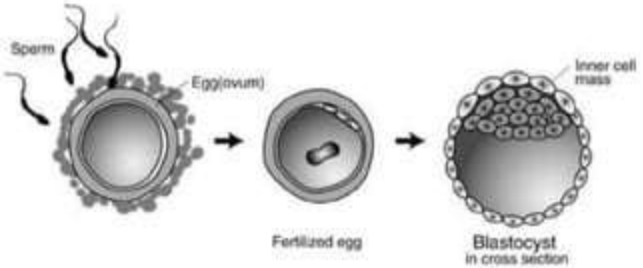
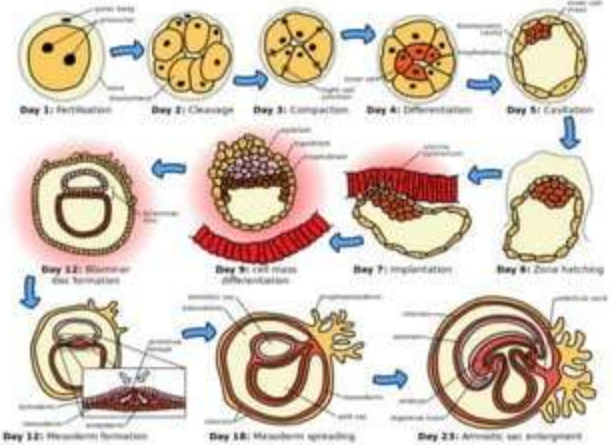
BACKGROUND

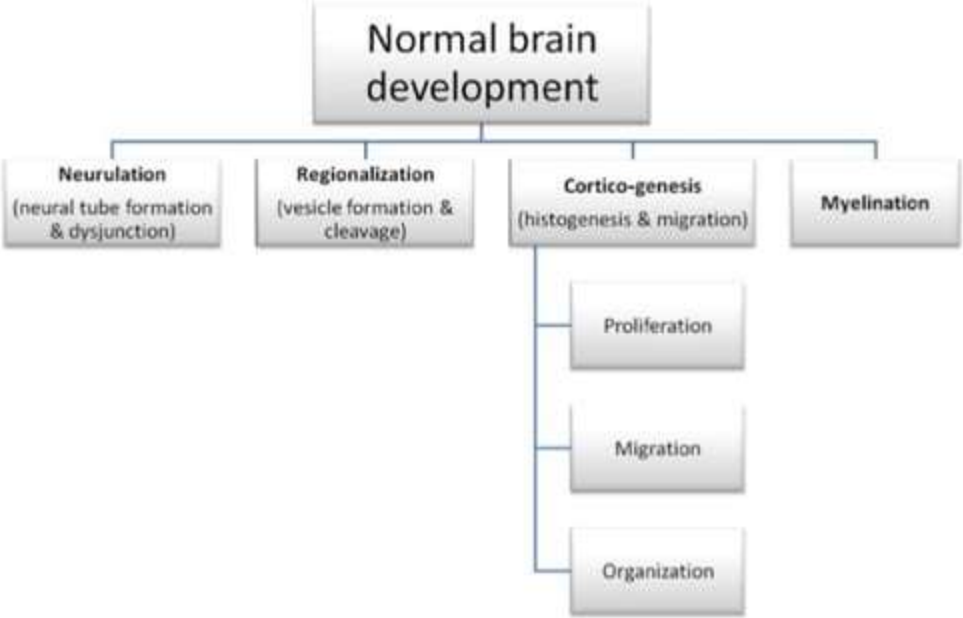
Meningoencephalocele:

- is a neural tube defect
- is PROTRUTION of intracranial structure outside the confined of the skull through the cranial bone defect, occurs MIDLINE
- Insidence 1: 5000 to 10.000 births worldwide
- Intracranial structure consists of brain tissue and cerebrospinal fluid
- Brain tissue might be nonfunctional glial elements or vital structure such as hypothalamus

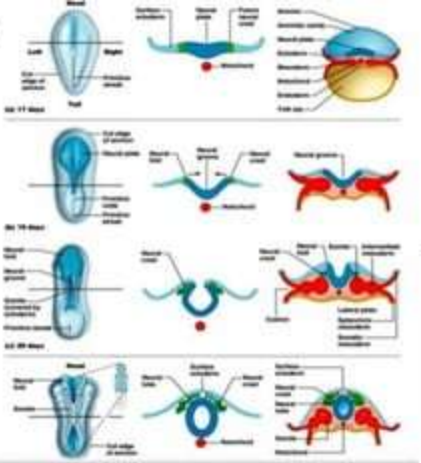
Moment of embryology



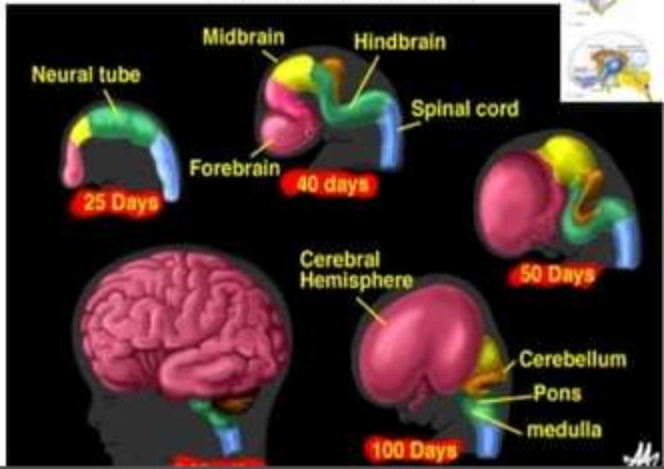


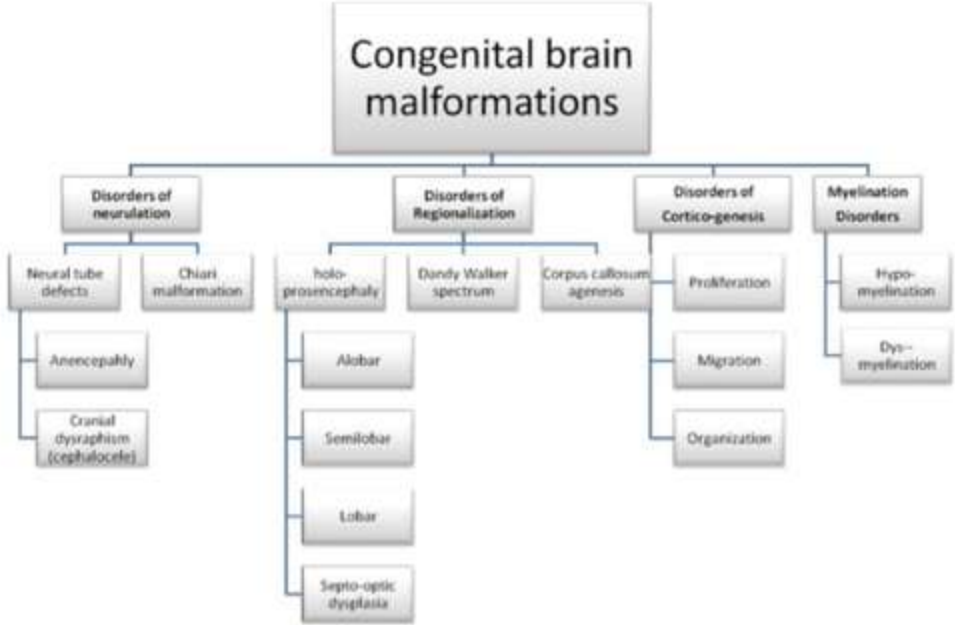

Normal brain development
Neurulation
(neural tube formation & dysjunction)
Regionalization
(vesicle formation & cleavage)
Cortico-genesis
(histogenesis & migration)
Myelination
Proliferation
Migration
Organization

Neurulation



Ventral induction

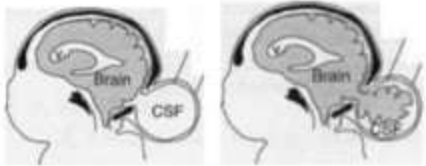



Congenital brain malformations
Disorders of neurulation
Neural tube defects
Chiari malformation
Anencepahly
Cranial dysraphism (cephalocele)
Disorders of Regionalization
holo-prosencephaly
Dandy Walker spectrum
Corpus callosum agenesis
Alobar
Semilobar
Lobar
Septo-optic dysplasia
Disorders of Cortico-genesis
Proliferation
Migration
Organization
Myelination Disorders
Hypo-myelination
Dys-myelination

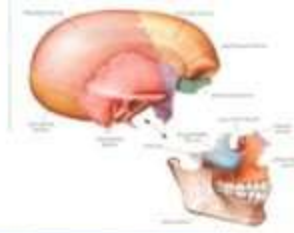
Cephaloceles

Pathological classification
(based on the contents of the sac)

Meningo-cele	Meningo-encephalocele	Meningo-encephalo-cystocele	Atretic cephalocele	Gliocele
CSF	CSF	CSF	Fibro-fatty tissue	CSF
	BRAIN (dysgentic & nonfunctioning)	BRAIN		Glial tissue
		VENTRICLES		

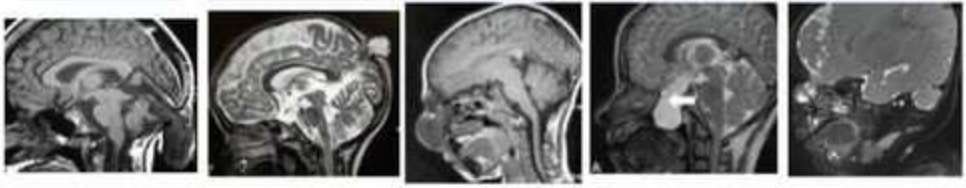


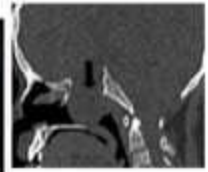
Cephaloceles



Anatomical classification
(based on the location of the defect)

Occipital	Parietal	Fronto-ethmoidal	Trans-sphenoidal	Nasal





Classification by. Suwanwela & Suwanwela*

Classification

System based on Suwanwela and Suwanwela[25]:

1. occipital: often involves vascular structures
2. cranial vault: comprises ≈ 80% of encephaloceles in Western hemisphere
   a) interfrontal
   b) anterior fontanelle
   c) interparietal: often involves vascular structures
   d) temporal
   e) posterior fontanelle
3. fronto-ethmoidal: AKA sincipital; 15% of encephaloceles; external opening into face in one of the following 3 regions:
   a) nasofrontal: external defect in the nasion
   b) naso-ethmoidal: defect between nasal bone and nasal cartilage
   c) naso-orbital: defect in the antero-inferior portion of medial orbital wall
4. basal: 1.5% of encephaloceles; (see below)
   a) transethmoidal: protrudes into nasal cavity through defect in cribriform plate
   b) spheno-ethmoidal: protrudes into posterior nasal cavity
   c) transsphenoidal: protrudes into sphenoid sinus or nasopharynx through patent craniopharyngeal canal (foramen cecum)
   d) fronto-sphenoidal or spheno-orbital: protrudes into orbit through superior orbital fissure
5. posterior fossa: usually contains cerebellar tissue and ventricular component

Suwanwela C, Suwanwela N. A Morphological Classification on Sincipital Encephalomenigoceles. J Neurosurg. 1972; 36:201-211

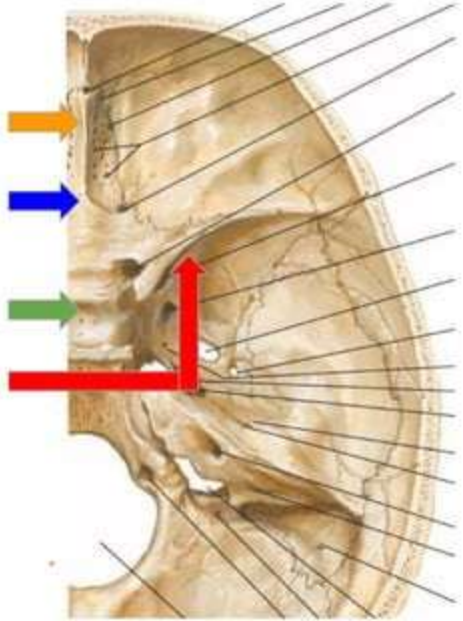
On Skull base the bone defect may be on:

ethmoidal bone: Protrude into nasal cavity through cribriform plate

spheno-ethmoid: Protrude into posterior nasal cavity

sphenoid bone: Protrude into sphenoid sinus or nasopharynx

fronto-sphenoid or spheno-orbital: Protrude into orbit



ETIOLOGY

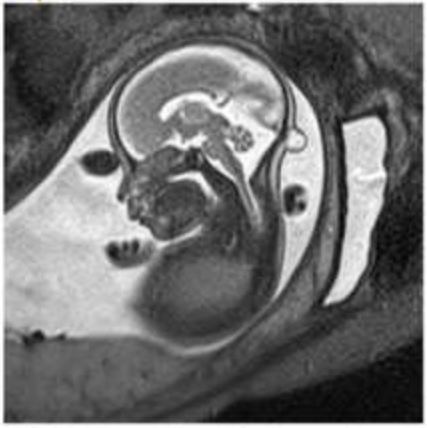
Two main theories*:

- Arrested closure of normal confining tissue allows herniation through persistent defect
- Early outgrowth of neural tissue prevents normal closure of cranial coverings

*Greenberg M., 2016. Developmental Anomalies in Handbook of Neurosurgery 8th ed. Thieme. New York. P255-256

DIAGNOSE

- Prenatal: Ultrasound, abnormal AFP maternal serum
- Birth: Visible mass, CT scan head (3D reconstruction), MRI head



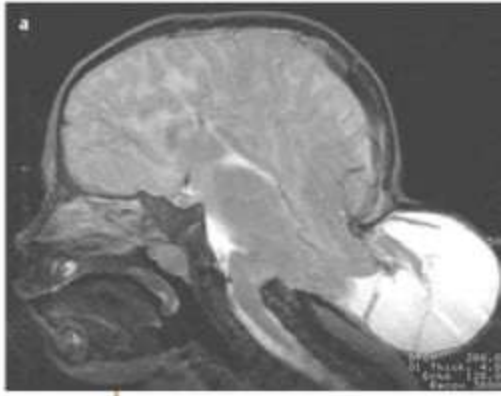
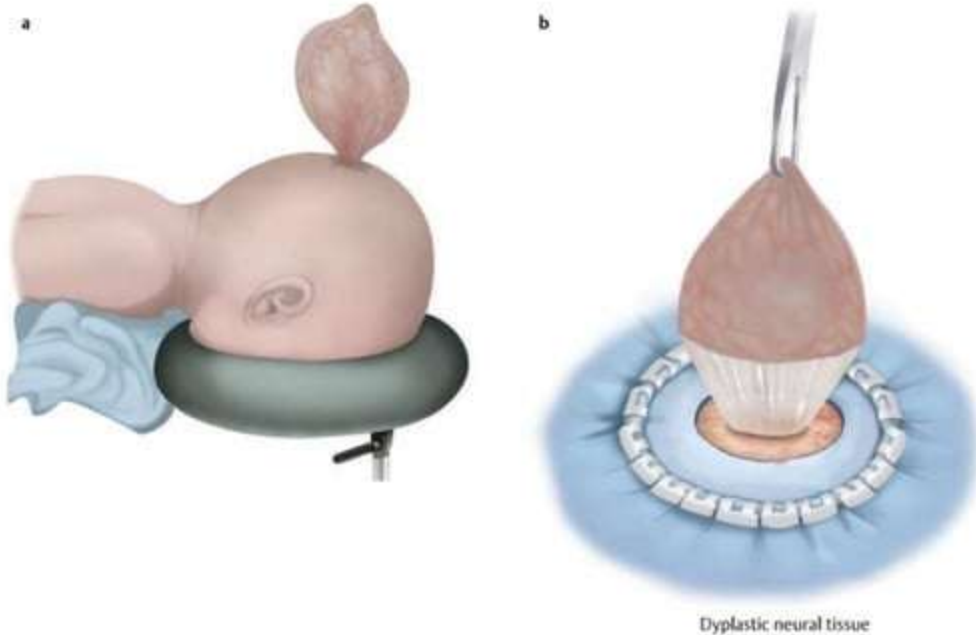


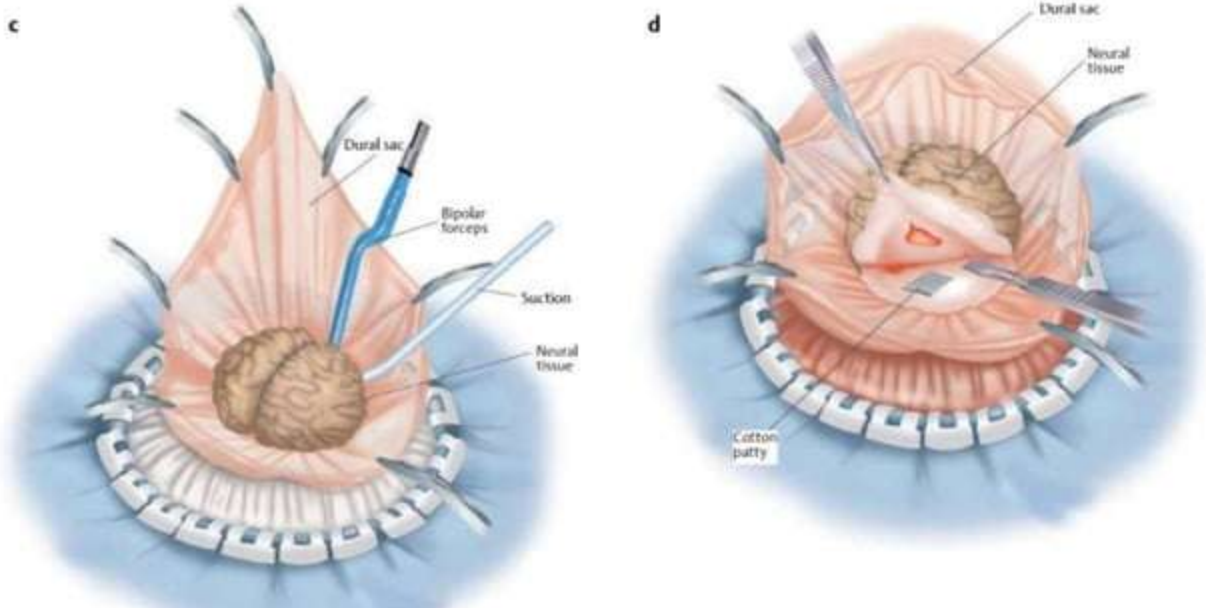
Fig. 22.3 Small occipital encephalocele seen on intrauterine magnetic resonance imaging (MRI).

MANAGEMENT

- Occipital encephalocele: SURGICAL EXCISION of the sac and water-tight dural closure. Hydrocephalus is often present and may need to be treated separately.
- Basal encephalocele: Transnasal approach



Drake JM, MacFarlane R. Encephalocele. in: Cheek W, ed. Arlas of Pediatric Neurosurgery. Philadelphia, PA: WB Saunders; 1996



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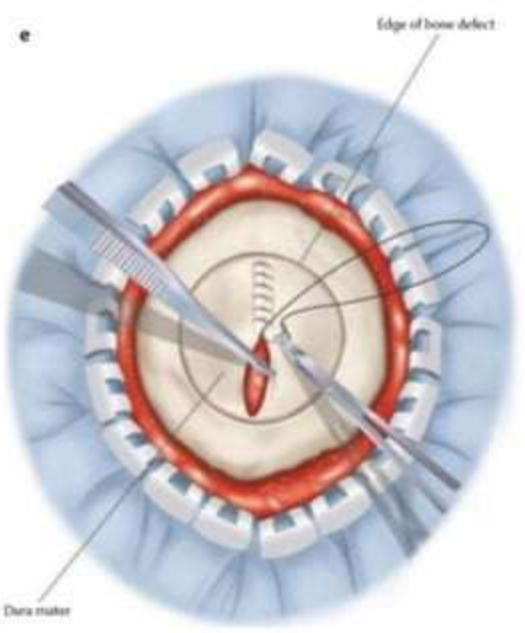

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OUTCOME

- Occipital encephalocele: The prognosis is better in occipital meningocele than in encephalocele.
- Prognosis is worse if a cerebral tissue is present in the sac, if the ventricles extend into the sac, or if there is hydrocephalus.
- Less than 5%of infants with encephalocele develop normally.

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THANK YOU