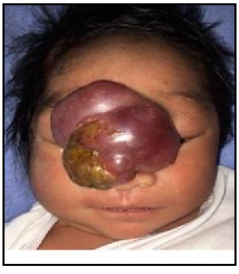
Indian Association of Pediatric Surgeons

Patient Information Sheet

ENCEPHALOCELE



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What is encephalocele ?

Encephalocele is a rare birth defect with sac-like protrusion from a bony defect in the skull. This is usually a skin covered sac which contains a part of the brain and the protective covering of brain (meninges). These are common at the back of head (occipital encephalocele), less common on forehead (fronto-nasal encephalocele) and very rarely on other sites. Encephaloceles containing brain tissue and meninges are termed as meningoencephalocele; those with brain tissue and spinal cord contents are termed as encephalomyeloceles; and those with only meninges are termed as cranial meningocele.

What causes this problem and how common is it ?

Encephalocele originates during the intra-uterine development of the fetus. When there is a defect in the developing nervous system, such that a neural tube, which give rise to brain and spinal cord, has improper folding and closing leading to outpouching of brain contents. The exact cause in not known. The incidence of encephalocele is reported to be 0.8-3 per 10,000 live births.

What are the symptoms ?

The sac-like protusion from the skull is the evident sign of encephaocele, which is noted at birth. Due to part of brain tissue in the encephalocele, there can be various neurological problems like increased head pressure due to obstruction of brain fluid circulation (hydrocephalus); smaller head size than expected for age (microcephaly); weakness in arms and legs (paraparesis or paraplegia); uncoordinated body movements (ataxia); delayed milestones; seizures; vision impairment; learning disability. Posterior encephaloceles are often symptomatic as the have protruding brain tissue in them; however anterior encephalocele mostly lacks brain tissue

contents, thus have better future outcome. Rarely, the encephalocele may be too small to get noticed. Encephalocele may be associated with various named syndrome like Fraser Syndrome, Roberts Syndrome, Warburg Syndrome etc.

When to see your doctor ?

After the birth, child should be seen by a pediatric surgeon for planning the elective surgery after assessment of the child.

How is it diagnosed ?

Encephalocele can be diagnosed on the antenatal ultrasound scan, as an out-pouching on head of the fetus. When encephalocele is identified antenatally or at birth, the initial ultrasound is followed by an detailed MR scan to delineate the extent and content of the encephalocele. Various associated neurological anomalies like Dandy-Walker malformation or Chiari Malformation should be identified during MR scan.

What are the treatments available ?

Surgical repair is the only option. Since encephalocele is a skin-covered lesion, there is no emergency to operate. Surgery should be planned any time after birth, after detailed assessment for associated conditions. Children with craniofacial abnormalities or hydrocephalous may be treated with additional surgeries.

Are there any alternatives to surgery ?

This lesion is precarious and the injury to the encephalocele can lead to grievous consequences, like rupture of encephalocele, exposure of brain tissue to the surrounding, infection of meninges and brain, and sudden death. Therefore, this lesion should be repaired. Till the child is planned for operation, the child should be nursed in lateral position with

help of cushions or use a cushioned ring to support the encephalocele. Children with functional disabilities, however, will require supportive treatment with the help of neurologist, child psychologist and physiotherapist.

What does the operation involve ?

The surgery involves either reduction of protruded brain contents back into brain cavity and covering the defect with the tough layer of meninges or may involve extending the hole on the skull by cutting the portion of skull (craniotomy), removal of nonfuntioning brain tissue and excess sac of encephalocele.

What are the possible complications / what happens after the operation ?

Child is usually started on feeds on the same day of surgery and is discharged within 2-3 days after surgery. Additional surgeries may be required for craniofacial, neurological abnormalities or hydrocephalus. Post-operative infections may occur in the wound or as meningitis. Hydrocephalus may also develop if not present before.

What is the outlook or future of these children ?

Surgical correction of encephalocele often does not worsen any pre-operative neurological status of the children. Therefore children with preserved neurological status can lead a symptom-free normal life. Children who have neurologic derangement will be benefitted by special support group. Families of these children needs a genetic counselling and birth planning strategies for future pregnancies. Mothers of children with encephalocele and other spinal defects are advised to start the intake of folic acid before next conception and continue the same during the pregnancy.