Basal encephalocele

The only group that does not produce a visible soft tissue mass. May present as Cerebrospinal fluid fistula or recurrent meningitis. May be associated with other craniofacial deformities, including: cleft lip, bifid nose, optic-nerve dysplasia, coloboma and microphthalmia, hypothalamic-pituitary dysfunction. Iniencephaly is characterized by defects around the foramen magnum, rachischisis and retrocollis. Most are stillborn, some survive up to age 17.

Basal encephalocele is the rarest form of encephalocele, and transsphenoidal encephaloceles (TSE) represent the rarest form of basal encephalocele.

Basal encephaloceles are often associated with other midline anomalies such as hypertelorism, broad nasal root, cleft lip, and cleft palate. Optic disc anomalies such as pallor, dysplasia, optic pit, coloboma, and megalopapilla have been reported to occur in patients with basal encephalocele

Caprioli and Lesser report a case of a child with a sphenoethmoidal encephalocele and morning glory syndrome of the optic nerve. The presence of such optic nerve anomalies with facial midline anomalies should alert the clinician to the possible presence of a basal encephalocele [1].

see Transethmoidal encephalocele.

1)
Caprioli J, Lesser RL. Basal encephalocele and morning glory syndrome. Br J Ophthalmol. 1983 Jun;67(6):349-51. PubMed PMID: 6849854; PubMed Central PMCID: PMC1040063.