the usual final stage of the aneurysm evolution, but rather an ongoing dynamic process that has potential for further growth and mass effect. Aneurysmal growth may be due to accumulation of thrombotic materials, recurrent intramural hemorrhage, or development of intrathrombotic capillary channels, which may, in turn, thrombose or bleed (6). The natural history of most untreated giant aneurysms is extremely dismal. The literature contains very few cases of complete spontaneous occlusion of partially thrombosed giant BA aneurysms (1,12,13). However, during recent decades, good outcomes after the treatment of ruptured and unruptured pediatric aneurysms have increased (5).

**REFERENCES**


1. Loevner LA, Ting TY, Hurst RW, Goldberg HI, Schut L.. Spontaneous thrombosis of a basilar artery traumatic aneurysm in a child. AJNR Am J Neuroradiol. 1998; 19: 386-368
2. Huang LT, Shih TY, Lui CC. Posterior cerebral artery aneurysm in a two year old girl. J Formos Med Assoc 1996; 95: 170-172
3. Benoit BG, Wortzman G. Traumatic cranial aneurysms, clinical features and natural history. J Neurol Neurosurg Psychiatry 1973; 36: 127-138
4. De Tella OI Jr, Crosera JF, Herculano MA, de Paiva Neto MA. Giant intracranial aneurysm in three years old boy: case report. Arq Neuropsiquiatr. 2006; 64: 530-533
5. Huang J, McGirt MJ, Gailloud P, Tamargo RJ. Intracranial aneurysms in the pediatric population: case series and literature review. Surg Neurol. 2005; 63: 424-432
6. Massimi L, Moret J, Tamburrini G, Di Rocco C. Dissecting giant vertebro-basilar aneurysms. Childs Nerv Syst. 2003; 19: 204-210
7. Wardlaw JM, White PM. The detection and management of unruptured intracranial aneurysms. Brain. 2000; 123: 205-221
8. Kasner SE, Liu GT, Galetta SL. Neuro-ophthalmologic aspects of aneurysms. Neuroimaging Clin N Am 1997; 7: 679-692
9. Andrews BT, Edwards MS, Gannon P. Acutely thrombosed aneurysm of the middle cerebral artery presenting as intracranial hemorrhage in a 3-year-old child. Case report. J Neurosurg 1984; 60: 1303-1307
10. Tanabe M, Inoue Y, Hori T. Spontaneous thrombosis of an aneurysm of the middle cerebral artery with subarachnoid hemorrhage in a 6- year- old child: case report. Neurol Res 1991; 13: 202-204
11. Nakstad P. Spontaneous occlusion of traumatic pericallosal aneurysm and pericallosal artery. Neuroradiology 1987; 29: 312
12. Kanaan I, Lasjaunias P, Coates R. The spectrum of intracranial aneurysms in pediatrics. Minim Invasive Neurosurg 1995; 38: 1-9
13. Maeda K, Usui M, Tsutsumi K, Iijima A. Spontaneous occlusion of a giant basilar tip aneurysm and a basilar artery due to the dissection of both structures: case report. Surg Neurol 1997; 48: 606-609


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## LETTER TO EDITOR (VIEWERS CHOICE)

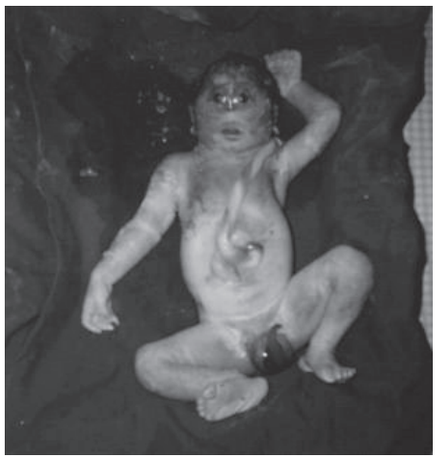
### AN UNUSUAL PRESENTATION OF CYCLOPIA WITH SITUS INVERSUS

*Shailesh R Singi*

A boy was born of non-consanguineous parents in our hospital to a 32 year old woman after 36 weeks of uneventful gestation. There was no history of exposure to any drugs or substances that might be considered teratogenic and there was no family history of dysmorphism. At birth the baby showed Cyclops malformation. He survived only few minutes. On examination there was a centrally placed orbital fossa, which was diamond shaped and contained a single eyeball with double corneas and pupils. A proboscis-like appendage was located above the eye on the lower part of the forehead. The infant was microcephalic; the head circumference was 20 cm. The mouth was triangular. The palate was high arched. The ears were low set and neck short (Fig. 1). He had hypospadias and liver palpable 3 cms below left coastal margin. An ultrasonography of abdomen and skull revealed features suggestive of complete situs inversus and holoprosencephaly (HPE) respectively. The postmortem examination could not be done.

Three cardinal features characterize Cyclopia -single orbital fossa with varying degree of fusion presenting one or two eyes, a proboscis-like appendage above and holoprosencephaly. (1) The Cyclops malformation is characterized by a single orbital fossa located in the area normally occupied by the bridge of nose. The socket is diamond shaped, with open angles extending beyond the globe. There may be all degrees of fusion, from two eyes in same orbital cavity, to a single globe with two corneas and two pupils to a single eye. There may be one or no optic nerve. There is a nose like appendage, called proboscis found above the eyeball. It

**Fig. 1: Newborn-Photograph illustrating two corneas, pupils and iris in single globe with triangular orbital fossa and rudimentary proboscis above the eye.**



may be covered with normal skin. The cyclopean variety is most severe of the holoprosencephalies or median faciocerebral dysplasias. In these disorders (also known as archinencephaly or alobar holoprosencaphaly) there usually is single ventricle along with absence of corpus callosum, chiasma and septum pellucidum. The thalami are fused. The second and fourth part of cranial nerve is absent. Other central nervous system abnormalities include microcephaly, or hydrocephaly occasionally, spina bifida, anterior encephalocele, acrania or anencephaly. There may be variety of oral manifestations. The mouth is generally triangular, ears are low set, malformed or absent. There may be cleft palate or bifid uvula. Cyclopia with unilateral or bilateral abnormality of the ear may be associated with agnathia and has been called cyclopia hypognathus. There may be also anomalies of lung, cryptorchidism or hypospadias (1).

Cyclopia is one clinical presentation of severe craniofacial anomalies resulting because of failure of division of telencephalon into two cerebral hemispheres with a single ventricle - Alobar holoprosencephaly (2,3). It is further subdivided into three types-pancakes, cup and ball depending upon degree of failure of rotation. In De Meyer's series of 23 cases of holoprosencephaly associated with major facial types 22 had alobar holoprosencephaly (3). Approximately 50% cases with holoprosencephaly are associated with cytogenetic abnormalities. At present, 12 loci on different chromosomes for HPE have been identified. These include sonic hedgehog gene (SHH) at 7q 36, ZIC2 at 13 q 32, SIX3 at 2q 21, and TGIF at 18p11.3, human DKK1, PTCH and TDGF1. Of these, mutations in SHH are the most frequently identified single gene defect associated with human holoprosencephaly. Mutation in SHH has been identified more frequently in familial holoprosencephaly (autosomal dominant) than sporadic holoprosencephaly (18% and 3.4% respectively). So far, a total of 46 mutations have been identified in 364 unrelated HPE patients, corresponding to mutation detection rate of 12.6%. Even with detection of mutation because of intrafamilial variability observed it might be difficult to predict the phenotype of fetus. At present ultrasound appears a very effective mode of prenatal diagnosis (4).

**REFERENCES**

1. Nyhan WL, Sakati NO. Single Syndromic Malformations: Cyclopia. In Genetic and malformation Syndromes in Clinical Medicine. Chicago, Year Book Publishers, Inc. 1977; 401-403.
2. Cohen MM Jr. Perspectives on holoprosencephaly: Part III. Spectra, distinctions, continuities, and discontinuities. Am J Med Genet. 1989; 34: 271-288.
3. Demyer W, Zeman W, Palmer CG. The Face Predicts the Brain: Diagnostic Significance of Median Facial Anomalies for Holoprosencephaly (Arhinencephaly). Pediatrics. 1964; 34: 256-263.
4. Thakur S, Singh R, Pradhan M, Phadke SR. Spectrum of holoprosencephaly. Indian J Pediatr 2004; 71: 593-597.

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