Cyclops Deformity In Benin City, Nigeria: A Case Report

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Citation

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Abstract

Cyclopes are rare congenital abnormalities; a severe form of holoprosencephaly resulting in children being born with just one eye. It results from failure of the cerebral hemisphere to separate during fetal development. The incidence is 1 in 13,000 live births but present in 1 in 2500 pregnancies that end up as miscarriage. It is incompatible with life. In this report we present a Cyclops delivered via cesarean section on account of ante partum hemorrhage secondary to placental previa type 11a.

INTRODUCTION

The term cyclopia is commonly used to describe either the abnormality of true cyclopia in which a single median eye is the only ocular structure present or synophthalmia in which two globes (right and left eyes) are partially fused in the median position. Cyclopia (also cyclocephaly or synophthalmia) is a rare and severe form of holoprosencephaly and is a congenital disorder (birth defect) characterized by the failure of the embryonic prosencephalon to properly divide the orbits of the eye into two cavities. Its incidence is 1 in 16,000 in born animals, and 1 in 250 in embryos.[1, 2,3]

Garzozi and Bankay reported that true cyclopia is a rare anomaly in which the organogenetic development of the two separate eyes is suppressed. A case of true cyclopia with normal karyotype was presented. There was a history of the use of an intrauterine device for contraception and of drug ingestion during early pregnancy. An anatomico-pathological study of the monster with detailed presentation of the cyclopean eye was also reported.[4, 5]

Typically, cyclopia present with the nose either missing or replaced with a non-functioning nose in the form of a proboscis. Such a proboscis generally appears above the central eye, and is characteristic of a form of cyclopia called rhinencephaly or rhinocephaly.[6]

Although cyclopia is very rare, several cyclopic human babies are preserved in medical museums (e.g. The Vrolik Museum, Amsterdam).[7] There are also two known cases of children with Down syndrome being born with one eye.[8] Some extreme cases of cyclopia have been documented in cats. In such cases, the nose and mouth fail to form, resulting in suffocation shortly after birth.[9]

Suggested risk factors include maternal diabetes, infections during pregnancy (syphilis, cytomegalovirus, toxoplasmosis, herpes and rubella), drugs taken during pregnancy (alcohol, aspirin, lithium, anticonvulsants, hormones, retinoic acid, anticancer agents and fertility drugs) and physical agents like ultra violet light, previous pregnancy loss and first trimester bleeding[10, 11, 12]

Genetic problems or toxins can cause problems in the embryonic forebrain-dividing process[13]. One highly teratogenic alkaloid toxin that can cause cyclopia is cyclopamine or 2-deoxyjervine, found in the plant Veratrum californicum (also known as corn lily or vetch weed). The mistake of ingesting Veratrum californicum while pregnant is often due to the fact that hellebore, a plant with which it is easily confused, is recommended as a natural treatment for vomiting, cramps, and poor circulation, three conditions that are quite common in pregnant women.[14]

In 2005, a kitten with cyclopia, “Cy”, was born in the United state and died about one day after birth.[15]

Cyclopia deformities have been recognized for centuries. Cyclops are mentioned in Greek mythology at least three times[16]. Homer described cyclopic giants who lived as shepherds in Sicily. Hesiods tells of three cyclopic storm gods. Brontes, Steropes and Arges, who were sons of Uranus and Gaea. They belonged to the family of Titans. A third group of cyclopean giants had abdominal arms. These mythical prehistoric workmen are said to have built the walls of Mycenae and Tiryns and were objects of worship because

of their strength. Cyclopean deformity were said to be caused by punishment from the gods for the wickedness of man.[16]

Familial occurrences in twins and in consanguineous marriages have been documented and would be consistent with a single gene abnormality[17]. In normal differentiation of the cephalic midline structures there is increase expression of Pax-2 gene and inhibition of Pax-6 gene from the notochord. However, inappropriate expression of these genes may result in cyclopia[18]. Mutation of sonic hedgehog gene (SSH) has also been implicated in the formation of cyclopia. Other genes that play a role in the formation of these defects include ZIC2, TGIF and SIX3[19]. In recent years several isolated case report of Cyclops or holoprosencephaly in humans and animals have been associated with abnormal chromosomes[20].

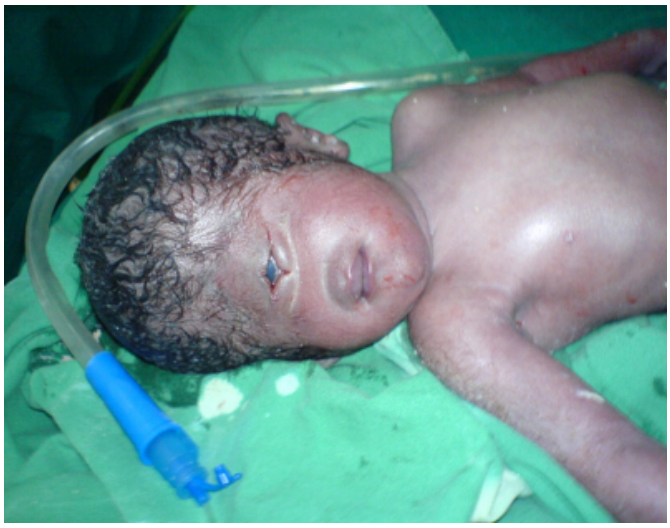
CASE REPORT

A live female infant (1.5kg) delivered by cesarean section at 01.35hours on 23rd August 2006 on account of Ante-partum Hemorrhage (APH) secondary to placenta previa type IIA with breach presentation in the labor ward theater of the University of Benin Teaching Hospital, Benin City Nigeria. Apgar score was three in one and two at five minutes. Multiple congenital abnormalities were apparent at birth (fig.1), single centrally located eye, absence of the nose, small mouth, and extra digit on the right upper phalanx with spinal bifida occulta. Pronounced dead ten minutes following delivery.

The 37year old mother who resides in Benin City was unbooked and has three normal, healthy living children; two females and one male.

Figure 1

Figure 1



POST MORTEM EXAMINATION

This 1.5kg female infant whose placenta weighed 0.3kg was 42.7cm long and the head circumference 24cm. Several facial bones especially those associated with the nose were missing. Cardiac anomalies were noted which included patent ductus arteriosus, patent foramen ovale, left ventricular hyperplasia and dilated left atrium.

A single umbilical artery was present which was connected to the right common iliac artery. An accessory spleen was present

The 69gm brain was holoprosencephalic and the cerebral hemispheres were not cleaved rostrally. Posterioly, there was a partial cleavage with formation of symmetrical occipital lobes. The brain stem and cerebellum appeared normal. The optic nerve and olfactory tract were not seen. Coronal section of the brain confirmed the presence of a holosphere.

MICROSCOPIC EXAMINATION

Microscopic examination showed cystic degeneration of the thyroid follicles. The ventricular cavities were not developed but the fourth ventricle and aqueduct of sylvius was identified.

DISCUSSION

Cyclops is usually associated with trisomy 13 or 18 and may be associated with other abnormal chromosomes abnormalities or occasionally with normal chromosomes. It arises early in gestational life causing lack of cleavage of the hemisphere into right and left cerebral hemisphere[20, 21]. As noted in the post mortem report above the cerebral

hemisphere of the reported female infant were not cleaved rostrally, but she had a partial posterior cleavage forming symmetrical occipital lobes. Its incidence is usually sporadic[22]. As at the time of this report, only two cases have been delivered since the inception of University of Benin Teaching Hospital, Benin city. However, this is the first case ever of two Cyclops and grossly different from the second case (a female with a proboscis above the only eye) yet to be reported.

The severity of the facial abnormality reflects the severity of the brain abnormalities to a certain extent[23]. This is exemplified by the post mortem report. The total systemic deformity associated with cyclopean abnormality are generally inconsistent with fetal viability and only a few cyclopic individuals survive pregnancy to be still born or die shortly after birth[24, 25]. One cyclops was reported to have lived for ten years[26]. This female infant died 10 minutes after delivery.

Multitudes of environmental agents (physical, chemical and biological) have produced experimental Cyclops because they alter chromosome numerically and structurally[27]. Cyclopia has been reported in children with diabetic mothers which supports the multifactor theory of cyclopia development because diabetes is a multifactor disease but this seems unlikely[28].The 37 year old mother was not diabetic and gave no history associated with the risk factors mentioned above.

CONCLUSION

At present all the facts concerning cyclopia are consistent with some chromosomal abnormality, even if chromosomal observation represents only one group of several etiologies. Cyclopia is a very rare anomaly which one may never have the opportunity in a lifetime to witness. This is the first case of cyclops to be reported in Benin City, Nigeria.

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References

1. Taber's Cyclopedic Medical Dictionary, ISBN 0-8036-0654-0
2. Gupta RC, Gupta VK, Gupta S. Human cyclopia with associated microstoma and and anencephaly. Indian J Ophthalmol 1981;29:121-3
3. Dubourg C, Bendavid C, Pasquier L, Henry C, Odent S,David V. Holoprosencephaly Orphanet J Rare Dis. 2007; 2: 8.
4. Garzozi HJ, Barkay S. Case of true cyclopia. Br.J.Ophthalmol. 1985 April v. 69(4): 307-311
5. Arathi N, Mahadevan A, Santosh V, Yasha TC, Shankar SK. Holoprosencephaly with cyclopia - Report of a pathological study . Neurol India 2003;51:279-82
6. Dark, Graham (2007). Rhinocephaly. In Online Medical Dictionary. Retrieved July 23, 2008.
7. "Vrolik Museum, Department Of Anatomy And Embryology, University Of Amsterdam".
8. Available @ www.messybeast.com/freak-face.htm".
9. Cyclopia. Wikipedia, the free encyclopedia. Available @ http://en.wikipedia.org/wiki/cy_(kitten).
10. Available @ www.medterms.com/script/main/art.asp?articlekey=15530".
11. Mollica F, Pavone L, Sorge G. Maternal drug ingestion and cyclopia. J Pediatr. 1981 Apr; 98(4):680.
12. Benawra R, Mangurten HH, Duffell DR. Cyclopia and other anomalies following maternal ingestion of salicylatesJ Pediatr.1980 Jun; 96(6):1069-1071.
13. Tapadia MD, Cordero DR, Helms JA. It's all in your head: new insights into craniofacial development and deformation. J Anat. 2005 November; 207(5): 461-477.
14. Veratrum californicum. Available @ "Teratology Society". http: // www.teratology.org/members/JMF presentation/tsld011.htm.
15. Petty, Terrence (January 11, 2006). "Not a Hoax, One-Eyed Kitten Had Bizarre condition" (HTML). Animaldomain. LiveScience.com. Retrieved on 2007-02-05.
16. Mondi R. "The Homeric Cyclopes. "Foktale, Tradition and Theme "Transaction of the American Philogical Association. 1983, Vol. 113; pp 17-38.
17. Ming JE, Muenke M. Diseases and Holoprosencephaly Am J Hum Genet. 2002 November; 71(5): 1017-1032.
18. Kim JW, Lemke G. Hedgehog-regulated localization of Vax2
controls eye development Genes Dev. 2006 October 15; 20(20): 2833-2847
19. Cordero D, Marcucio R, Hu D,Gaffield W, Tapadia M,Helms JA. Temporal perturbations in sonic hedgehog signaling elicit the spectrum of holoprosencephaly phenotypes J Clin Invest. 2004 August 16; 114(4): 485-494.
20. R.O. Howard, Chromosomal abnormalities associated with cyclopia and synophthalmia. Trans Am Ophthalmol Soc. 1977, 75: 505-538.
21. Chan A, Lakshminrusimha S, Heffner R, Gonzalez-Fernandez F. Histogenesis of retinal dysplasia in trisomy 13 Diagn Pathol. 2007; 2: 48.
22. Cotran RS, Kumar V, Robbins SL; Robbins Pathologic Basis of Diseases 5th edition, Philadephia, W.B. Saunder 1994; pp 1302.
23. Poirer J et al. manual of basic micropathology. Philadephia: Saunders 1990; pp 199.
24. Lilard R: The cyclops deformity a case report. Rocky Mt. Med. J. 1964; 61:32.
25. Khudr G., Olding L: Cyclopia AM. J. Dis Child. 1973; 125:120.
26. Vare AM: Cyclopic AM J. Ophthamol. 1973; 75:880.
27. Pei W, Williams PH, Clark MD, Stemple DL, Feldman B. Environmental and Genetic Modifiers of squint Penetrance during Zebrafish Embryogenesis. Dev Biol.2007 August 15; 308(2): 368-378.
28. Dekabeu A, Maee K: Occurrence of Neurological abnormalities in infants of diabetic mothers. Neurology. 1956; 8:193.

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