JOJ Ophthalmology
ISSN: 2473-5477

Juniper Publishers
key to the Researchers

Review Article
Volume 10 Issue 5 - April 2024
DOI: 10.19080/JOJO.2024.10.555800

JOJ Ophthalmol
Copyright © All rights are reserved by Ndiaye JMM

The Cyclopie: About an Observation and Review of Literature

Ndiaye JMM*, Mendy GK, Sow AS, Badji MD, Attye M, Diallo R and Ndoye Roth PA

Ophthalmology Department of Aristide Le Dantec Hospital, Dakar, Senegal

Submission: March 02, 2024; **Published:** April 17, 2024

***Corresponding author:** Ndiaye JMM, Ophthalmology Department of Aristide Le Dantec Hospital, Dakar, Senegal

Summary

Cyclops are the most severe form of holoprosencephaly. It is a rare polygenetic malformation that occurs during early embryonic development. It is a single eye, or by extension, two eyes, contained in the same median orbit. We report a case of cyclopy at the Aristide Le Dantec National University Hospital. It was premature of 35 SA + 05 days, born of a pregnancy complicated by eclampsia, without pathological history in the parents. A prenatal ultrasound had objectified a severe hydro-amnios with a microcephaly. The ophthalmological examination showed a single median orbital cavity containing two different sized eyeballs. The general examination objectified a microcephaly, an arhinia and an abscess of the right foot. The orbito-cerebral CT showed a major cerebral malformation with microcrania type and a cyclopia. The child died after 27 days of ectopic life from respiratory distress. Cyclopia is a non-viable malformation whose prenatal diagnosis is based on the realization of an obstetric ultrasound from 12eSA. An etiological assessment is essential, it makes it possible to look for environmental and genetic risk factors.

Keywords: Cyclopies; Holoproencephalon; Polymalformation

Introduction

Cyclopia is the most severe form of holoprosencephaly (HPE), which is the most common congenital defect in the human anterior brain. Cyclopy represents 2% of HPE cases [1,2]. It is a rare polygenetic brain malformation that results from a defect in median cleavage of prosencephalon to right and left hemispheres and occurs during early embryonic development (between the 18th and 28th gestation days) [3,4]. Incompatible with postnatal life, cyclopia can be diagnosed on ultrasound of the first trimester, which can allow early termination of pregnancy [5]. We report an observation about cyclopie at the Aristide Le Dantec National University Hospital.

Observation

It was a premature born by cesarean section at 35 SA + 05 days that was referred to us by the maternity hospital for the management of a polymalfomative cephalic syndrome. The 35-year-old mother had no particular pathological history. There were no defects or malformations in the siblings. During pregnancy, she received three prenatal consultations. An obstetric ultrasound performed at 32 SA, revealed severe hydramnios and a suspicion of microcephaly. No sequelae of infection were noted (HIV1, HIH2, Syphilis and Hepatitis B). At birth, the Apgar score was 7/10 at the 1st minute and 8/10 at the 5th minute. The weight was 1555 g, the height 33cm and the cranial perimeter was 22 cm. The examination found a microcrania, a single central orbit, containing two different sized eyeballs. The right eye was more voluminous, seat of a megalocornea and a corneal xerosis. There was bilateral arhinia, small ears normally implanted, and an abscess in the right foot (Figure 1). There was no cleft palate or malformations of the extremities. The neck, spine and long bones were continuous and regular.

Cerebral computed tomography at J17 of life objectified an orbital anomaly with a single orbit and two eyeballs of normal size and appearance separated by a median partition (Figure 2). It revealed a major cerebral malformation, microcrania-cyclopy type, marked by an enlargement of the fourth ventricle and prepontic and quadrigeminal cisterns, an elevation of the cerebellum tent, a brain stem of normal density, a difficult individualisation of the central grey nuclei. There was a fluid density of the «cerebral hemispheres» apart from the persistence of the cerebral parenchyma in the frontal lobes punctuated by micro-calcifications. The lateral ventricles and the third ventricle were not individualized. The diagnosis of synophthalmia or incomplete cyclopia (or cyclopia imperfecta) was retained. Therapeutic management consisted essentially of resuscitation means such as hydro-electrolytic rehydration, continuous oxygen

therapy, and blood transfusion. Ophthalmological treatment was administered based on eye washing, tobramycin eye drops, dexpanthenol (corneal scarring). Treatment of the right foot abscess was a parenteral antibiotic based on Cefotaxime 100mg/d and Amikacin 20mg/d. The evolution was marked by multiple episodes of respiratory distress of progressive aggravation. The last, occurred at J27 of life led to the death of the newborn.

Figure 1: Anterior view of the newborn with both eyes in a single orbit (white arrows), arhinia (black arrow) and absence of proboscis.

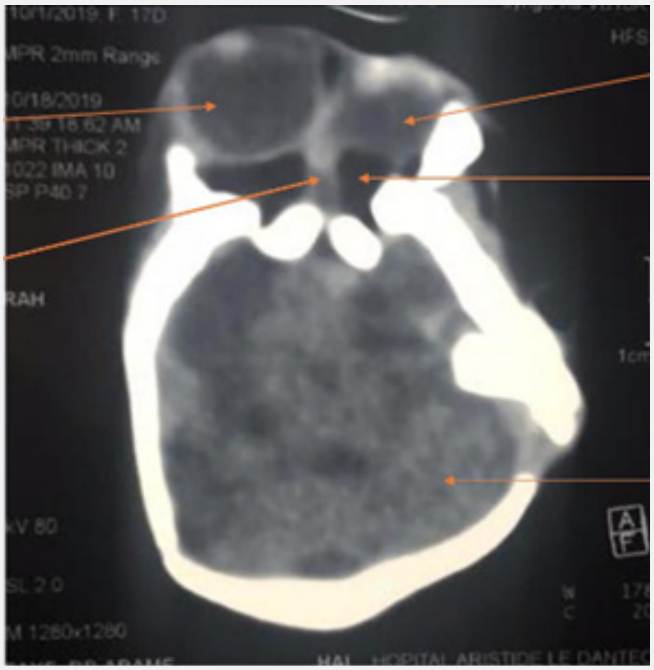

Figure 2: Cerebral CT showing cyclopia with a single orbit and two fused eyeballs.

How to cite this article: Ndiaye JMM, Mendy GK, Sow AS, Badji MD, Attye M, et al. The Cyclopie: About an Observation and Review of Literature. JOJ Ophthalmol. 2024; 10(5): 555800. DOI: 10.19080/JOJO.2024.09.555800

Discussion

The term cyclopy is commonly used to describe either the anomaly of true cyclopy in which a single median eye is the only ocular structure present or synophthalmia in which two globes are partially fused in the middle position [6]. A third type of ocular deformation was observed in cyclopia, which is the total absence of eyeballs (anophthalmia) [7]. About 1.05 in 100,000 births, including stillbirths, are identified as cyclopeans. It is the second most common malformation of the human brain after anencephaly [8].

Epidemiological Aspects

In the Very Rare Anomaly study of the International Organization for Surveillance and Research on Congenital Anomalies, 20 monitoring programs in 25 countries provided data on cyclopia over a period of study from 1968 to 2006. A total of 257 infants with cyclopia were identified among the 25,580,661 births, resulting in a total prevalence of 1.0 per 100,000 births.

Genetic Aspects

Several genes have been involved in holoprosencephaly, but its genetic basis remains obscure. Chromosomal abnormalities are not specific. They are numerical or structural and can concern any chromosome [10]. Trisomy 13 is the aneuploidy most frequently associated with cyclopia. Trisomies 18 and 21 were also described, as well as triploidism [9]. Cyclopia is a heterogeneous condition, which may result from chromosomal abnormalities, genetic mutations, or teratogenic environmental factors [11].

Risk Factors

The etiology of HPE is very heterogeneous. Besides the genetic causes, this pathology can be caused by environmental or metabolic factors. However, few epidemiological studies have had a sufficient sample to examine risk factors. The only formally recognized environmental or maternal factors are insulin-dependent diabetes, well known to increase the risk of fetal malformations, and maternal alcoholism with a cumulative risk from smoking [11]. Fenugreek, which is contraindicated during pregnancy, is among the oldest medicinal and condiment plants used for anti-inflammatory properties. It is very popular with women in the Mediterranean, and for a few years in Senegal. In Morocco, it has been mentioned anecdotally in the genesis of prosencephalic induction abnormalities and more significantly in neural tube malformations [12]. Inbreeding, as a risk factor, has also been reported.

Ophthalmological Aspects

Three types of cyclopia are distinguished: monophthalmia, synophthalmia with two fused eyeballs and anophthalmia [14]. The newborn of our observation presented synophthalmia without proboscis. Prenatal ultrasound remains the best way for early diagnosis. In the first two trimesters, with careful examination of the face, the diagnosis of cyclopia is possible. In the third trimester, she found severe hydramnios, highlighting the signs of HPE, cyclopy with or without proboscis and arrhinia [15]. Very advantageous fetal MRI can be performed reliably from 18 weeks of gestation. Pathological examination helps to find an eye with different degrees of duplication of intrinsic ocular structures called synophthalmia, with a single optic nerve without chiasma. On histological examination, different abnormalities can be discovered such as retinal dysplasia, chorioretinian coloboma and optic nerve hypoplasia. The muscles of the eye are usually imperfectly separated and located around the optic nerve in conoid form [16].

Prognosis - Survival

Cyclopia often leads to miscarriage or stillbirth. Survival after birth is usually a few hours. A very limited survival, rarely exceeding the first week of life, has been reported. Thus, the particularly long-life span of our child cyclops, who survived 27 days, is an exception.

Conclusion

Holoprosencephaly is a non-viable malformation whose prenatal diagnosis is based on the realization of an obstetric ultrasound from the 12eSA. Etiological assessment is essential, it makes it possible to look for environmental and genetic risk factors, hence the interest of genetic studies in the prenatal assessment.

References

1. Monuki ES (2009) Forebrain Development: Holoprosencephaly (HPE). Neurosci Academic Press, USA, Pp; 303-313.
2. Winter TC, Kennedy AM, Woodward PJ (2015) Holoprosencephaly: A Survey of the Entity, with Embryology and Fetal Imaging. Radiographics 35(1): 275-290.
3. High Authority Of Health (HAS) (2018) Holoprosencephaly (HPE) & related forms. Chronic disease guide. Saint-Denis La Plaine, pp. 49.
4. Dupé V, Dubourg C, de Tayrac M, David V (2017) From cyclops to reality: a fresh look at the genetics of holoprosencephaly. Med Sci 33(11): 924-926.
5. Măluţan AM, Dudea M, Ciortea R, Mureşan M, Bucuri CE, et al. (2017) Cyclopia and proboscis - the extreme end of holoprosencephaly. Rom J Morphol Embryol 58(4): 1555-1559.
6. P Otuaga, A Eweka, A Oni, L Ebite (2007) Cyclops Deformity In Benin City, Nigeria: A Case Report. The Internet J Neurol 10(1).
7. Rodrigues LD, Arakeri SU, Dwarampudi RS (2019) Rare association of cyclopia with craniospinal rachischisis. Indian J Pathol Microbiol 62(2): 283-286.
8. Sarnat HB, Resch L, Flores-Sarnat L, Yu W (2014) Precocious synapses in 13.5-week fetal holoprosencephalic cortex and cyclopean retina. Brain Dev 36(6): 463-471.

How to cite this article: Ndiaye JMM, Mendy GK, Sow AS, Badji MD, Attye M, et al. The Cyclopie: About an Observation and Review of Literature. JOJ Ophthalmol. 2024; 10(5): 555800. DOI: 10.19080/JOJO.2024.09.555800

9. Rioli O, Amar IM, Bakker E, Bermejo-**Sánchez** MK, Bianchi E et al. (2011) Cyclopia: an epidemiologic study in a large dataset from the International Clearinghouse of Birth Defects Surveillance and Research. American J med genetics. Seminars in medical genetics 157(4): 344-357.

10. Tekendo-Ngongang C, Muenke M, Kruszka P (2020) Holoprosencephaly Overview. In: Adam MP, Ardinger HH, Pagon RA, et al. Gene Reviews editors. Seattle, pp. 75.

11. Blaas H-GK (2018) Holoprosencephaly. Obstetric Imaging: Fetal Diagnosis and Care (Second Edition): Elsevier pp. 204.

12. Seddiki A, Messaouidi S, Amrani R (2017) The role of fenugreek in the occurrence of neural tube closure anomaly: a warning signal from Morocco. Herbal medicine 15(3): 155-158.

13. Fatnassi R, Turki E, Belhaj J, Labidi I, Ben Regaya L, et al. (2011) Holoprosencephaly: etiopathogenesis and phenotypic characteristics. About four observations. Morphol 95(310): 79-82.

14. Deftereou TE, Tsoulopoulos V, Alexiadis G, Papadopoulos E, Chouridou E, et al. (2013) Congenital disorder of true cyclopia with polydactylia: case report and review of the literature. Clini exp obs gynecol 40(3): 460-462.

15. Hsu TY, Chang SY, Ou CY, Chen ZH, Tsai WL, et al. (2001) First trimester diagnosis of holoprosencephaly and cyclopia with triploidy by transvaginal three-dimensional ultrasonography. European J Obst Gynecol Reproduct Biol (96): 235-237.

16. Fallet-Bianco C (2018) Neuropathology of holoprosencephaly. American J med genetics 178(2): 214-228.

This work is licensed under Creative Commons Attribution 4.0 License
DOI: 10.19080/JOJO.2024.09.555800

Your next submission with Juniper Publishers will reach you the below assets

- Quality Editorial service
- Swift Peer Review
- Reprints availability
- E-prints Service
- Manuscript Podcast for convenient understanding
- Global attainment for your research
- Manuscript accessibility in different formats
  (Pdf, E-pub, Full Text, Audio)
- Unceasing customer service

Track the below URL for one-step submission
https://juniperpublishers.com/online-submission.php

How to cite this article: Ndiaye JMM, Mendy GK, Sow AS, Badji MD, Attye M, et al. The Cyclopie: About an Observation and Review of Literature. JOJ Ophthalmol. 2024; 10(5): 555800. DOI: 10.19080/JOJO.2024.09.555800