

# Congenital Heterochromia Iridis in a Nigerian Girl Child

Omolase Charles Oluwole, A. K. Akinwalere, O. A. Adeosun, B. O. Omolase, M. Y. Majekodunmi

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See end of article for  
authors affiliations

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Correspondence to:  
Omolase Charles Oluwole  
Federal Medical Centre  
PMB 1053,Owo,Ondo State  
Nigeria

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This report is that of a six month old Nigerian girl child with complete heterochromia iridis. There was no associated hypo-pigmentation of her skin or hair. There is no history of similar occurrence in her family. The early presentation of the child may be due to the fact that the parents are enlightened. Cycloplegic refraction done did not reveal any significant refractive error. However we intend to place the patient on coloured contact lens in the nearest future to reduce chromatic aberration to the barest minimum and to conceal the hypo pigmented iris most especially in view of the fact that the patient is a girl child. Though congenital heterochromia iridis appears rare in our population, there is need to educate the general population about this ocular condition so that they can be more receptive to affected individuals.

**H**eterochromia iridis is an ocular condition in which there is difference in the colour of the irides of the two eyes or where part of one iris has a different colour from the remainder. It may be complete heterochromia iridis or partial/sectoral heterochromia iridis. Partial or sectoral heterochromia is less common than complete heterochromia iridis and it is typically found in autosomally inherited disorders such as Hirschprung's disease and Waardenburg's syndrome. Heterochromia iridis arises from relative excess or lack of pigment within an iris or part of an iris which may be genetically inherited or due to mosaicism or acquired by disease/injury<sup>1</sup>. Congenital heterochromia iridis is usually inherited as an autosomal trait. The colour of the iris is determined by the concentration and distribution of melanin pigments within the iris tissues<sup>2,3</sup>, thus any alteration in these factors may result in difference in colour of the iris<sup>2</sup>. The affected eye may be hyper pigmented (hyperchromic) or hypo pigmented (hypochromic). The conditions that could make the iris darker (hyperchromic) include Lisch nodules, ocular melanosis, oculodermal melanocytosis (naevus of Ota)<sup>4</sup>, pigment dispersion syndrome, Sturge - Weber

syndrome characterized by a port wine stain naevus in the distribution of trigeminal nerve, neurologic signs and angioma of the choroid often with secondary glaucoma<sup>5,6</sup>.

Hypochromia iridis can arise from simple heterochromia iridis which is characterised by absence of other ocular or systemic problems. Other causes include congenital Horner's syndrome and Waardenburg's syndrome<sup>7</sup>. Other conditions that are associated with hypochromia iridis are Piebaldism, Hirshsprung disease, Incontinentia pigmenti, Parry -Romberg syndrome .Acquired heterochromia iridis may also be due to injury, inflammation, use of certain eye drops or tumours. Acquired causes of hyperchromia iridis include siderosis, use of eye drops (prostaglandin analogues), melanoma of the iris, irido-corneal endothelium syndrome and iris ectropion syndrome<sup>4</sup>. The acquired causes of abnormally lighter iris are Fuchs' heterochromic iridocyclitis, acquired Horner's syndrome, chronic iritis, juvenile xantho-granuloma, leukaemia and lymphoma<sup>4</sup>. Central heterochromia iridis is an eye condition in which there are two different colours in the same iris.

In view of the rarity of congenital heterochromia iridis in our population, we decided to report this case so as to draw the attention of the people to this ocular condition.

### CASE REPORT

A six month old Nigerian female child presented to the Eye Clinic of Federal Medical Center, Owo, Ondo State, Nigeria in September, 2010 on account of discolouration of both eyes since birth. There was no other associated ocular complaint. There was no history of maternal illness in the course of her pregnancy. She was delivered at 35 weeks of gestation through emergency caesarean section at the University Teaching Hospital, Ado Ekiti, Nigeria. The indication for the caesarean section was ante-partum haemorrhage and transverse lie. She had neonatal jaundice at the third week of life for which exchange blood transfusion was done. She was diagnosed of having Glucose 6-phosphate dehydrogenase (G6PD) deficiency at the hospital. The patient is the last born of four children in a monogamous setting. Both parents are school teachers. There is no history of similar occurrence in her family.

Examination of the patient revealed that she was yet to attain neck control at six months of age. There was no area of hypo-pigmentation on her skin or head (no white forelock on forehead). Ocular examination revealed visual acuity of light tracking in both eyes. There was complete hypochromia iridis in both eyes and the pupils were briskly reactive. However anterior segment examination did not reveal any other abnormality. Dilated pupil funduscopy revealed pink optic disc with a cup-disc ratio of 0.3 and normal vessels in both eyes. There was mild hypopigmentation of the retina. The macula appeared normal. The patient had cycloplegic refraction with the aid of gutt atropine but there was no significant refractive error thus no glasses were recommended. The patient was also seen by the paediatrician in view of delayed developmental mile stones. The patient was to be seen periodically at the Eye Clinic and she was to be placed on coloured contact lens later.

### DISCUSSION

The human iris can have different colours. There are three true colours in the iris that determine the outward appearance; brown, yellow and grey. How much of each colour a person has, determines the appearance of his or her eye colour.<sup>8</sup> Eye colour is a polygenic trait and it is determined by the amount of

melanin in the iris. Blue iris are due to lack of melanin while brown eyes indicate richness in melanin in the iris.<sup>9</sup> People who have dark hair and skin tend to have higher levels of melanin resulting in brown iris. However people with lighter skin and hair tend to have lower level of melanin which makes their iris lighter. People with heterochromia iridis tend to have chromatic aberration which could cloud their vision. This was the reason why we decided to refract the patient reported with a view of correcting any refractive error that could have been detected. The decision to request that the child be brought for periodic review was to appropriately manage the spherical aberration which is likely to be marked when the child is older. The need for the child to use coloured contact lens when she is old enough cannot be overemphasized. The coloured contact lens is expected to reduce chromatic aberration to the barest minimum and also help to conceal the hypopigmented iris. This measure was discussed with the mother and she accepted the measure based on the information she was given. The relative early presentation of the child is appreciated and is likely to be due to the fact that the parents are enlightened and well motivated. It is also likely to be due to the fact that the parents were bothered about the cosmetic challenge of hypochromia iridis most especially in a girl child. It is the considered opinion of the authors that the child may not derive pride from the striking appearance conferred by hypochromia iridis in the nearest future. There are few reports of heterochromia iridis in Nigeria. The author for correspondence reported a case of simple heterochromia iridis in a 15 year old Nigerian girl<sup>9</sup>. There was no associated systemic or ocular abnormality in the patient<sup>9</sup>. Onabolu also reported two cases of Waardenburg's syndrome in a Nigerian family<sup>10</sup>. Both patients had white forelock, heterochromia iridis and sensory-neural deafness.<sup>10</sup> Amoni et al reported two cases of Waardenburg's syndrome in Nigeria.<sup>11</sup> A Japanese review of 11 albino children with Waardenburg syndrome found that all had sectoral / partial heterochromia iridis.<sup>12</sup>

### CONCLUSION

Though congenital heterochromia iridis appears rare in our population, it is important to adequately address the ocular challenges associated with the condition. It is important to continue to motivate the parents of this child so that she can receive adequate care. The populace should also be educated about this ocular condition so that they can be more receptive.



**Fig 1:** Girl child with bilateral heterochromia iridis

#### Author's affiliation

Dr. Omolase Charles Oluwole  
Department of Ophthalmology  
Federal Medical Centre, Owo  
Ondo State, Nigeria

Dr. A. K. Akinwalere  
Department of Ophthalmology  
Federal Medical Centre, Owo  
Ondo State, Nigeria

O. A. Adeosun  
Department of Ophthalmology  
Federal Medical Centre, Owo  
Ondo State, Nigeria

B. O. Omolase  
Department of Radiology  
Federal Medical Centre, Owo  
Ondo State, Nigeria

M.Y. Majekodunmi  
Department of Ophthalmology  
Federal Medical Centre, Owo  
Ondo State, Nigeria

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#### Glaucoma

Comparing the advantages and disadvantages of bleb dependent glaucoma surgical techniques which have been performed for more than a century with the recent bleb independent techniques, Trabeculectomy in its various modified forms is still the choicest procedure.

**M Lateef Chaudhry**  
Editor-in-Chief