J-MWAN 3 (2014) 23 - 34

Original Article

CAREGIVER BURDEN IN NIGERIAN PARENTS OF CHILDREN WITH OCULOCUTANEOUS ALBINISM

Authors: **Dr. Frances OA Ajose FRCP (Lond), Dr Olufolakemi M Cole MBBS**
Department of Medicine, Lagos State University Teaching Hospital,
Ikeja, Nigeria.
Corresponding: Dr Frances OAAjose
Department of Medicine
P.O. Box 1723 Surulere, Lagos, Nigeria.
francesajose@yahoo.co.uk
Tel: +2348038559066

ABSTRACT

Introduction

Oculocutaneous albinism (OCA) is a rare recessively inherited abnormality of pigmentation, worldwide in distribution, but with particularly devastating consequences in the African. The child is usually born healthy with Caucasoid phenotype to unsuspecting normally pigmented African parents. Uncomplimentary myths about aetiology and characteristics of albinos, the lack of protective skin pigment in the harsh African sun and poor vision combine to limit education, recreation and occupation of OCA There is no cure or treatment for OCA but strategies directed at sun avoidance and protection can very effectively prevent skin cancers and low vision aids can improve school performance. To be effective, these interventions need to be started in infancy. In the absence of any national medical or social support for the OCA, the burden of care has been thrust on unguided, ill-equipped parents with dire consequences.

Objectives

Our study sought to explore the caregiving burden of Nigerian parents of OCA to identify their support needs, determine the appropriate interventions to guide Nigerian policy makers.

Method

By means of interviewer assisted semi- structured questionnaire we explored the objective and subjective parameters of the care-giver burden of 76 consenting parents of OCA who attended monthly Albino Patient Support meetings at the Lagos State University Teaching Hospital, Nigeria over a 4-year period.

Results
Caregiver burden of parents caring for children with OCA is very heavy financially and psychologically. Predictors of caregiving success are parental education with above average income status.

Conclusion.
Unmet needs of parents include medical, educational and social support.

Keywords: albinism, African, caregiver burden, social support, education .

Introduction

Oculocutaneous albinism (OCA) [1,] is one of a heterogeneous group of rare recessively inherited abnormalities of pigmentation, worldwide in distribution, but with particularly devastating consequences in the African[2,3]. The two main types are ocular albinism (OA)[4] which affects only the eyes and Oculocutaneous albinism (OCA) which affects the eyes, skin and hair. At least fourteen genes[5] have been identified with up to 500 mutations, found to be associated with the various subtypes of albinism.**The primary abnormality**[6] **is the mutation of one of several genes** that code the instruction for making one of the many proteins necessary for the production of melanin[7], the chemical that gives color to the skin, hair and eyes.

The function of melanin include prevention of sunburn and skin cancer by filtering ultraviolet radiation (UVR)[8] entering the skin as well as maturation of the fovea of the retina in the eye[9]. Poor fovea development leads to ocular abnormalities that include strabismus, reduced stereoscopic vision and nystagmus from infancy, and reduced visual acuity that range from 20/40(6/12) up to 20/200(6/60.)[9]These visual abnormalities confer varying degrees of functional blindness that may significantly limit education, occupation and recreation.

Albinism is found in all races and ethnic groups with a worldwide frequency of about 1: 17,000 and a carrier rate of 1:70.[1]There is however, wide regional variation in prevalence due to the effect of founder mutation and incidence of consanguineous marriages. OCA occurs in 1:10,000 African Americans but only 1:36,000 white Americans.[1] within Africa, frequency ranges from as low as 1:15,000 in Eastern Nigeria[10,11] to as high as 1:1000 in the Tonga tribe of Zimbabwe.[12]

The African oculocutaneous albino (OCA) has yellow to ivory colored hair instead of the black African hair but the hair retains the African kinky hair texture. The skin color ranges from desirable brown African complexion to very white Caucasoid skin without any pigment. The pupils also lack pigment and may be grey or blue. In addition, the eyes are often crossed (strabismus) with constant rapid eye movements (nystagmus) and associated photophobia. This phenotype makes the African albino very different from his peers and very conspicuous in any gathering.
The OCA infant is otherwise a healthy child born without any structural deformity, to unsuspecting normally pigmented African parents who are often perplexed by it. Uncomplimentary myths[10, 13]abound about aetiology and characteristics of albinism which predispose to postnatal psychological distress in the new parents. As the child grows, the psychological impact of looking different as well as rude and disparaging jests[10,13]about albinism cause psychological distress in the child which may be shared by the parents. The harsh African sun predisposes to sun intolerance and distressing sun burns[3] the management of which most parents are unequipped. Effect of poor vision[14] is often first noted when the child repeatedly runs into stationary objects and later at school when educational performance appears not to match the child's intellect [15]or when the child performs clumsily at ball games.

There is no cure or treatment[1] for oculocutaneous albinism neither are there effective camouflage options. However strategies directed at sun avoidance and protection[16] can very effectively prevent sun burns and skin cancers and low vision aids[17] can improve school performance.

To be effective, these interventions to preserve the skin health and vision of the OCA need to be started very early in life[16]. Therefore the parents of the OCA child must be conversant with appropriate sun protection strategies right from postnatal ward. Unfortunately this does not appear to be the case in view of the large population of young Nigerian OCA seen on the streets without any consideration for sun protection whatsoever, with many of them walking around with advanced skin cancers[10]. Teachers have an important role to play in the educational success of the child because the OCA child requires considerable classroom support[16] to minimise the effect of short- sightedness, again this does not appear to be so, as too many albinos are unable to complete secondary education in spite of adequate intellectual ability[15].

A heavy burden of care therefore rests on the parents of an OCA child for their physical, educational and emotional development. However, Nigeria is yet to determine the special medical, educational or social consideration to be provided for families with OCA as there is really no research-based knowledge to guide. We found no study that even remotely addressed the needs of the carers of the OCA child. Our study therefore sought to explore the perspectives of Nigerian parents caring for OCA children.

Objectives

To identify the unmet needs of parents of children with OCA, determine the appropriate interventions to support the parents and provide this information to Nigerian policy makers.

Method

By means of interviewer assisted semi-structured questionnaires we explored the objective and subjective parameters of the care-giver burden of 76 consenting parents of OCA children who attended the monthly Albino Patient Support Meetings (APSM) at the Lagos State University Teaching Hospital, Nigeria between 2009 and 2013.

The 5-part questionnaire comprised record of demographic data and socio economic status of the parents; details of the support received from health care providers and school teachers at various levels; parents' assessment of the physical and developmental burden of albinism on childcare; parents' assessment of their emotional and social burden as caregiver of their OCA child while part 5 evaluated their adaptive strategies and unmet needs.

As we found no previous study on this subject, our questionnaire was specially designed with help from available caregiver assessment tools[18]. Response format was by 4-point Likert type scale.The questionnaire was administered by FOA at the parents' first meeting and repeated six months later to assess the benefit of the APSM.
The data was analysed by simple descriptive statistics using SPSS 17.
FOA conceived the study and conducted the interviews, while OMC collated and analysed the data. The study was approved by the LASUTH Research Ethics Committee.

RESULT

The parents were 25 fathers, 48 mothers and 3 elder siblings aged 25- 56years. 25% had more than one albino children including 5 sets of discordant twins. 40% were in monogamous families, 32% were polygamous and 29% separated, divorced or unmarried. 60% of the babies were born at government health facilities in various parts of the country, 29% at private or church facilities and 11% at traditional birth attendant units. 13% of parents were in the high income bracket while 33% were medium, 46% low income and 8% were wholly dependent on charity at time of interview. 42% of parents completed primary education, 40% secondary, while 12% had tertiary education and 7% were professionals but 6% had no formal education. **(Table1)**

Immediate postnatal reaction

96% of mothers were caught unawares at the birth of their first albino child due to unknown family history. 87% admitted being perplexed. A short-lived urge to abandon the baby at the healthcare facility was entertained by 5% and one parent actually did but returned within 24 hours to claim the child. Positive attitude of the fathers and in-laws significantly relieved the tension in the mothers.

Healthcare and School support

All parents lamented the dearth of information from healthcare workers at all levels. Often the labour room nurses were as perplexed as the parents and in a few cases were visibly scared at the sight of the emerging white haired newborn baby. Paradoxically, only the traditional birth attendants offered any form of advice at birth of the OCA and they even attempted targeted skin care procedure to the new born in the form of vegetable oil baths and body rinse with herbal infusion known to contain polyphenols and antioxidants[19].

Total helplessness about the health requirement of the child persisted in 80% until their first attendance at the APSM. Average age of the OCA at their parents' first attendance at APSM was 9 years while the adult OCA unaccompanied by parents were on average 23 years.

Only 5% of OCA received appropriate school support in the form of classroom setting, learning aids and personalised notes mostly from the more expensive private schools. Most teachers were reported to be unaware of the special needs of the OCA either for sun protection or vision support, and made no effort to assist. In such cases the OCA would normally drop out of school.

Physical and developmental burden

Although the children generally enjoyed good health, and parents appreciate their often pleasant demeanour, artistry and philosophical intellect, the albinism increased the cost of child care by 300-400% in most families. This consisted of expensive topical sun screens, sun protection apparels that were trendy enough to be acceptable by children and discourage jeers from peers. Other expenses include costly indoor games to distract OCA from outdoor sports, and baby-sitting arrangement in infancy.

Coping with the occasional moody spells of the OCA is an area most parents admit incompetence and this stems from the paradox of restraining a healthy boisterous lad or lass from following their peers out to play in bright sunshine.

Poor school performance due to low vision was the most distressing objective and subjective stressor and onset of skin cancer the most psychologically devastating parameter. There were more school drop outs among the lower income group. Skin care was also poorer in this group as they had more advanced solar elastosis (pre-cancers).

Economic burden

Parents in the higher income bracket admitted spending as much as 3 to 4 times more on the OCA than other children. Parents considered the OCA the more fragile child and were more likely to send the OCA to fee-paying private kindergarten and primary schools whilst the other children attended free government schools. More money was spent in custom made school uniforms necessary to cover arms and legs. Vision aids and other educational support tools normally offered freely to the blind, were not given free to albinos. Some parents lost income when they had to suspend their open air market trading till after the OCA started school because the open air environment was found to be intolerable by the OCA.

Emotional and Social burden

'There is no hiding place for the mother of the albino' is the traditional saying that captures the social life of the OCA parent. She is known by all in the community and in the market. In fact the entire family of the OCA are tagged with the OCA as 'mother of the european, 'father of the european', 'sister, brother, e.t.c., of the european'. These family members are expected to respond with cheers to such appellation as some favours may even be attached. Paradoxically the jests smeared at the OCA themselves are not always complimentary especially by the youth who nevertheless meant no harm. Examples are 'poor sighted european,' 'the one who must not eat salt,' etc. Some siblings resorted to fighting the community youths in defence of the OCA,

Parents admitted *loss of personal liberty* in spending more parenting time for the OCA for baby sitting at home to enforce sun avoidance. The OCA also required additional time piloting to and from school long after other children has outgrown such monitoring.

Less than 15% of parents rated their parenting of their OCA as successful before attending the APSM but within 6 months nearly 60% had considered their parenting significantly improved.

We found that predictors of caregiving success were parental education and above average family income.

DISCUSSION

The African child with OCA is a healthy child physically and mentally[20] and loved by parents but the challenges posed by sun sensitivity, low vision and cultural beliefs require home based interventions that must be started in infancy. Parents are unprepared for this task because the birth of an OCA child is not usually anticipated. Most parents are totally ignorant about the correct nature of albinism or where OCA comes from until they give birth to one, which often is not a pleasant experience initially. Unfortunately, healthcare professionals appear to be no better informed than the parents, and from labour ward to school age, more than 80% of our cohort of parents of OCA received no formal guidance regarding the special needs of these children. Therefore the parents of OCA, left uninformed and unguided, are exposed to informal, often misleading, directives from anyone anywhere. This is a likely contributor to the gaping difference between the lot of OCA in Africa and elsewhere. It is insightful that until their first attendance at APSM, up to 90% of parents of OCA had no clue about sun protection, yet skin damage from unprotected exposure to the tropical sun in infancy and childhood is the seed that produces skin cancers by teenage. Similarly, the fact that poor vision slows down the learning and performance of OCA is not adequately addressed in schools leading to frustrated OCA and their parents and large cases of school drop outs among the OCA[10].

Although caregiving is the natural responsibility of a parent, some children like the OCA, have needs beyond the normal that require institutional support of the parents. Health workers in maternity and under-five clinics do not appear to understand their vintage position in the prevention of skin cancer in the OCA, just as the teachers are largely ignorant of their role in the educational outcome of the OCA pupil. The parents are expected to be knowledgeable about sun avoidance and protection measures. They must be aware of sun-sensitizing toiletries and medicaments[21] that may increase the OCA's predisposition to sunburn. Parents are also expected to understand the limitation in the vision of the OCA and available aids and compensatory mechanisms. Most importantly, parents must be prepared to help the child through periods of emotional distress when the psychological impact of looking different[22] becomes overwhelming for the child. None of these skills is innate and must be learned.

Predictors of caregiving success in our families with OCA children appear to be parental education and above average economic status. This may be due to educated parents' ability to source from elsewhere, information that is not obtained from healthcare providers, to assist them in caring for their children. Parents with good economic status can afford to procure and enforce the sun protection items like sunscreens, hats and wearing apparels. They are also likely to provide shielded vehicular school transportation, vision aids, special print books, and supplement the teacher's capability to help the child. For children born to less privileged families, healthcare providers' lack of support and neglect of educational support may be responsible for the poorer outcome of the OCA from these homes.

It is important that parents of the OCA be equipped to provide required educational[16] and emotional support[23] from the day the child is born and reinforced at immunisation and under-five clinics and at every opportunity thereafter. Educational establishments with OCA pupils or students must understand the sun protection requirements which may include appropriate modification of school uniforms to minimise skin exposure, and exclusion from activities that involve prolonged unprotected outdoor exposure. Similarly to be considered are strategies to reduce the effect of the OCA short-sightedness on learning ability[16], that include seat location in class, special assistance with note taking and extra time for written examinations.

Most societies including children story writers[24] are obsessed with appearance and those with a visible difference like the African OCA are usually devalued. Therefore sun protection and vision aids alone do not fix the problems of the OCA. The parents must also receive appropriate social support and be equipped with skills to build coping strategies[25]in their children. These include development of emotional resilience and initiative-taking strategies as well asappropriate vocabulary to use in responding to curious or cruel comments. These skills must be learned in social support systems and are highly cost-effective considering the enormous cost[26] to society of caring for skin cancer, school drop outs, and maladjusted youths.

Society must be rehabilitated[27] to reduce its prejudice of the OCA the school, being the child's first environment, must take appropriate steps to educate both staff and students about albinism and discourage unkind utterances and gestures from fellow students. While discouraging outdoor games for the boisterous and youthful OCA appropriate indoor games must be substituted that will help to develop confidence, social skills and resiliency whilst improving cognitive and academic ability. Learning to enjoy indoor games early in life is a tool for lifelong stress management[28] for the OCA who must forever be in the shade.

Conclusion

Our study, the first to exclusively evaluate caregiver burden of parents caring for children with OCA, found the burden of care on Nigerian parents to be very heavy both financially and psychologically. Predictors of caregiving success are parental education with above average income status. Their unmet needs include medical, educational and social support.

Demographics of caregivers of children with Oculocutaneous albinism

Relationship of care giver		
Father	25	32.9%
Mother	48	63.2%
Elder sibling	3	3.9%
Age range		
<30yrs	15	19.7%
30-40	49	64.5%
>40	12	15.7%
Sex		
Male	26	34.2%
Female	50	65.8%
Marital status		
Monogamous	30	39.4%
Polygamous	24	31.6.%
Divorced/separated/never married	22	28.9%
Health facility where OCA was born		
Government	46	60.5%
Private/ Religious	22	28.9%
Traditional Birth Attendants	8	10.5%
Combined Family Income		
High	10	13.2%
Middle	25	32.9%
Low	35	46.1%
No regular income	6	7.9%

Parents Educational attainment		
No formal education	5	6.6%
Primary	32	42.1%
Completed Secondary	30	39.5%
Tertiary	15	19.7%
Professional	12	15.7%
Source of Info on albinism		
Premarital		
Labour ward	NIL	100%
Immunisation Clinics	NIL	100%
Under-5 clinics	3	3.9%
Attending Clinician	5	6.5%
Childs's school teacher	6	7.9%
Books and Monographs	5	6.5%
Albino support Group	4	5.2%
Public Media Programs	72	95%
Neighbours/Casual	25	33%
encounters/Passers by	22	29%

REFERENCES

1. Grønskov K, Ek J, Brondum-Nielsen K. Oculocutaneous albinism. Orphanet J Rare Dis. 2007; Nov 2;2:43.
2. HingES, Zeeb H, RepacholiMH: Albinism in Africaas apublic health issue. BMC Public Health 2006; 6:212.
3. Luande J, Henschke CI, Mohammed N: The Tanzanian human albino skin; Natural history. Cancer Apr: 15 1985;55vol 6:1823-8.
4. Summers CG. Albinism: Classification, clinical characteristics, and recent findings. Optom Vis Sci. Jun 2009;86 vol 6:659-62 doi: 10.1097/OPX.0b013e3181a5254c.
5. Moltó E, Fernández A, Phillips C, et al Albinochip: a universal genetic diagnosis for all known mutations associated to albinism. Oral communication, CS23 / C112 Official XXIst International Pigment Cell Conference - 21-24 Sept 2011, Bordeaux – France updated: September 04 2011 website www.ifpcs.org/.../programme accessed 06 Aug. 2014.
6. Zühlke C, Stell A, Käsmann-Kellner B. Genetics of oculocutaneous albinism Ophthalmologe. 2007 Aug; 104(8):674-80.
7. Brenner M, Hearing VJ. The Protective Role of Melanin Against UV Damage in Human Skin Photochem Photobiol. 2008; 84(3): 539–549.
8. Wright C.Y; The impact of solar ultraviolet radiation on human health in sub-Saharan Africa. South African Journal of Science, 2012;108(11-12): p. 45-51.
9. Creel DJ, Summers CG; King RA; Visual anomalies associated with albinism. Ophthalmic Paediatr Genet. 1990; 11(3):193-200 (ISSN: 0167-6784)
10. Okoro AN: Albinism in Nigeria. Brit J Dermatology 1975; 92:485-492
11. King RA, Creel D, Cervenka J, Okoro AN, Witcop CJ: Albinism in Nigeria with delineation of new recessive oculocutaneous type. Clin Genet 1980; 17 vol 4: 259-270.
12. Lund PM, Puri N, Durham-Pierre D, King RA, et al; Oculocutaneous albinism in an isolated Tonga community in Zimbabwe. Journal of Medical Genetics 1997;34:733-735.
13. Cordrin Arsene. The Albino Paradox – faith, culture, and despair in contemporary Tanzania; Africa Politics Portal Feb 2009: accessed 28 January 2013
14. Traboulsi EI, Green WR, O'Donnel FE Jr.: The eye in albinism. In Duane's Clinical Ophthalmology. Vol 4. Lippincott-Raven; 1999;1-20.
15. Fulcher T, O'Keefe M, Bowell R, Lanigan B, Burke T, et al: Intellectual and educational attainment in albinism. J Pediatr Ophthalmol & Strabismus. Nov-Dec 1995; 32 vol 6:368-72.

16. Lund PM, Gaigher R, A health intervention programme for children with albinism at a special school in South Africa. Health Educ. Res. 2002;17 (3): 365-372. doi: 10.1093/her/17.3.365
17. Yahalom C, Tzur V, Blumenfeld A, Greifner G, Eli D, et al. Refractive profile in oculocutaneous albinism and its correlation with final visual outcome. Br J Ophthalmol. Apr 1996 (4):537-9. doi: 10.1136/bjophthalmol-2011-300072. Epub 2011 Dec 1.
18. Selected caregiver assessment measures: a resource inventory for practitioners 2nd Edition 2012, at https://caregiver.org/caregiver/.../SelCGAssmt accessed on 16th August 2014
19. Ajose FOA. Nigerian Plants of Dermatological importance. Int J Dermatol 2007 Oct;46 Suppl 1:48-55
20. Ajose FOA, Parker RA, Merrall ELC, AdewuyaAO, Zachariah MP Quantification and comparison of psychiatric distress in African patients with albinism and vitiligo: a 5-year prospective study. J EurAcad Dermatol Venereol 2014 Jul 23;28(7):925-32. Epub 2013 Jul 23.
21. O'Gorman SM, Murphy GM.Photosensitizing medications and photocarcinogenesis. Photodermatol Photoimmunol Photomed. 2014 Feb;30(1):8-14. doi: 10.1111/phpp.12085. Epub 2013 Dec 3.
22. Landsdown, R., Rumsey, N., Bradbury, E., Carr, T., & Partridge, J., 1997; Visibly Different: Coping with disfigurement. Oxford: Butterworth Heinemann.
23. McGrouther D A. Facial disfigurement The last bastion of discrimination in the Psychology ofAppearanceBMJ 1997; 314:991
24. Jowett, S., & Ryan, T, Skin disease and handicap: an analysis of the impact of skin conditions. Social Science and Medicine,1985;20, pp.425-9.
25. Piliavin, I., Piliavin, J., & Rodin, L. Costs, diffusion and the stigmatised victim. Journal of Personality and Social Psychology,1976;32, pp.429-38.
26. Ademiluyi SA, Ijaduola GT; Occurrence and recurrence of basal cell carcinoma of the head and neck in Negroid and albinoid Africans. J Laryngol Otol, 1987;101(12): p. 1324-8.
27. Hearst D. Can't they like me as I am? Psychological interventions for children and young people with congenital disfigurement. Dev Neurorehabil 2007; 10 (2), pp.105-12.
28. Cobb Social support as a moderator of life stress. Psychosomatic Medicine1976;38, pp.300-314.