

A health intervention programme for children with albinism at a special school in South Africa

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Abstract

The genetic condition albinism has a high frequency among the Sotho people of northern South Africa. Affected children have pale hair, eyes and skin—a dramatic contrast to the normal dark pigmentation. Their visual performance is poor and many attend special schools for the visually impaired. Children with albinism experience problems that are, on the one hand, physiological, and, on the other, social-psychological and educational in nature. In this self-report study 38 children at a rural special school described their eye and skin problems, a direct result of their lack of pigmentation, as well as strategies they adopted to manage their condition. A further section of the study deals with the social adaptation difficulties experienced by these children. The questionnaire tested for local belief systems about albinism and how these impact on the socialization of children with albinism. The intervention strategy proposed in this study is based on the assumption that any attempt to address both the health and social problems should be of a holistic, interactionist nature, and be based on the values and belief systems of the local community. In addressing the physical problems, the proposed intervention programme focuses on sensible sun protection habits from a young age and the active parti-

cipation of the children. To alleviate the social problems a team (interactionist) approach including children, teachers, parents, health officials and the wider community is recommended.

Introduction

Albinism encompasses a group of inherited disorders of melanin synthesis [reviewed in (Oetting *et al.*, 1996)]. One type, called oculocutaneous albinism (OCA) as it affects the hair, skin and eyes, is prevalent among indigenous people in southern Africa (Kromberg and Jenkins, 1982; Lund, 1996) at a frequency of about 1:4000. Those affected have pale cream to chalky coloured skin, grey or hazel eyes and sandy to yellow coloured hair—a striking contrast in appearance compared with the normal dark pigmentation. This difference in appearance creates socialization and adaptation problems for the child with albinism.

As people with OCA have little protective melanin pigment their skin is extremely sensitive to sunburn, skin lesions and cancers, especially in Africa (Kromberg *et al.*, 1989; Lookingbill *et al.*, 1995). The lack of pigment also results in misrouting of the optic nerve fibres during development, leading to lifelong visual problems [reviewed in (Biswas and Lloyd, 1999)]. These problems include poor visual acuity, large refractive errors, rapid eye movements and a lack of binocular vision. Their extreme sensitivity to sunlight is an additional factor that contributes to the physical and social isolation of children with albinism as they are limited in the extent to which they can take part in outdoor games and activities. In

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addition to being constrained in terms of having 'fun', this also has a negative impact on the development of self-esteem obtained through the dynamics of group and team activities.

Most babies with albinism are born to normally pigmented parents. A single gene is involved in OCA2, the predominant type in sub-Saharan Africa. The product of the OCA2 gene is a protein found in the membrane of the organelles where melanin pigment is produced, whereas the OCA1 gene codes for the key enzyme in the synthesis of the pigment melanin. In OCA1 type albinism, if the enzyme is missing or non-functional, no pigment is produced, resulting in a total lack of pigment in the hair, skin and eyes. OCA2 children have inherited two non-working or defective forms of this gene, one from each parent. In addition to carrying this defective copy, each parent also has one normal copy, allowing for pigment production. There is a 25% chance that two carrier parents will have an affected baby.

This study investigates the health, education, social interactions and sun protection habits of 38 children pupils with albinism attending a special school for the visually impaired in northern South Africa, where the incidence of albinism is particularly high, at 1:1500 neonates (Venter *et al.*, 1995). The pupil's perceptions and knowledge about albinism were investigated, and their needs identified, with the aim of formulating an appropriate comprehensive health intervention strategy. Any health strategy or intervention programme should not only be geared towards alleviating the skin and eye problems so prevalent among people with albinism, but should also consider the social and psychological problems resulting from the striking difference in appearance between normally pigmented (black) people and those with albinism. In African communities, where traditional explanations concentrate on the misfortune of illness or deviance rather than on a biomedical explanation of its physical causes, such 'deviant' appearances can impact negatively on the socialization of a child with albinism. Intervention programmes that work effectively in western societies cannot be duplicated in non-western societies without taking

into consideration those cultural values that influence perceptions and actions about health and disease.

Subjects and method

The special school where this study was conducted recruits primary school children with visual impairment from a wide rural area of the northern province of South Africa. It is near the Tropic of Capricorn at an altitude of 1230 m, with an average of 8.5 h of sunshine per day, leading to high levels of ultraviolet (UV) radiation throughout the year. The school is well resourced, with a disabilities officer and nursing sister on site.

Of the 131 pupils at the school, 112 (85.5%) had albinism. Children in grade 5 (their sixth year at school) and above completed a written questionnaire (in English with a translator available if required) detailing socio-demographic, educational and health information, as well as protective measures adopted to manage their condition. In this study, only children old enough to express their views in English were included. Other methods, such as the 'write-and-draw' technique described by Hughes *et al.* and McWhirter *et al.*, may be appropriate for obtaining similar information, particularly on sun protection, from the younger age group (Hughes *et al.*, 1996; McWhirter *et al.*, 2000). The questions included both multiple choice and open style questions, with an opportunity for pupils to comment or ask questions at the end. The topics were chosen from previous similar surveys conducted in Zimbabwe (Lund, 1998, 2001).

The participants were asked about their perception of albinism and its causes. Each pupil was examined on a sunny day in autumn, to record cases of sunburn, facial skin lesions and the type of clothing worn. The skin creams used were investigated and the classroom environment noted.

All 38 pupils (18 males and 20 females; age range 11–16 years, mean 13.0 years) volunteered to participate. Most (32 pupils; 84.2%) belonged to the northern Sotho ethnic group. The number of children per family varied from one to 13, with

a mean of 5.3. About half (21 pupils; 55.3%) had no siblings with albinism, whereas 17 (44.7%) had at least one affected sibling. Twenty-four subjects (63.2%) had other close relatives with albinism. There was one case of twin girls with albinism in the study group.

Results

Appearance of pupils with albinism

All the pupils in this study, with the exception of one girl, had the type of albinism most frequently encountered in Africa, OCA2, with pale, chalky coloured skin, grey or hazel eyes and sandy to yellow hair. The girl had a different, rarer type called brown albinism. She produced more pigment than the others, having light brown skin, hair and eyes.

Skin and eye problems associated with albinism and their management

On examination all pupils with OCA2 were found to have dry, chapped and cracked lips, and 34 (89.5%) had sunburn. All wore hats protecting their upper but not the lower part of their faces. Two males had facial lesions that were being treated by the nurse. The three OCA2 pupils without sunburn were the only ones to wear thick clothing that protected their necks. The girl with brown albinism had no sun induced skin damage.

Most (32; 84.2%) respondents said their skin turned red and blistered when exposed to the sun and 21 (55.3%) reported having had skin sores. Other problems included chapped and painful lips and dry skin. Table I lists the precautions pupils took to protect their skin from sun. The creams used by 26 pupils were examined; half were inexpensive aqueous creams, with no sun-protection factor (SPF) and half had SPF values of 5–30.

Thirty-three (86.8%) pupils complained of poor eyesight and 30 (79.0%) of extreme sensitivity to light and sun. Visual acuity measurements varied from 6/60 (10% of normal vision) to 6/18 (33%) (M. J. Ika, pers. commun.). Although most had been for an eye test, few wore either spectacles or sunglasses, as shown in Table I. The few who had

Table I. Protective measures practised by schoolchildren with albinism (n = 38)

Measure	No. of pupils (%)
Sun protection	
applying cream	27 (71.1)
wearing hats	38 (100)
wearing long skirts or trousers	28 (73.7)
staying indoors/in the shade	13 (34.2)
wearing sunglasses	8 (21.1)
Eye care	
eye test	36 (94.7)
wearing spectacles	7 (18.4)

spectacles did not wear them all the time; none wore theirs to complete the questionnaire.

Special education for pupils with albinism

The classrooms in the school where the study was undertaken were specially adapted to reduce glare, with shutters on the windows at one side and a covered walkway on the other. Rooms were all equipped with a mobile blackboard on wheels that could be moved around to the best position at different times of day. The children were allowed to walk up to the board to read. All these measures helped to alleviate the poor vision and extreme sensitivity to sunlight experienced by those with albinism.

Sixteen (42.1%) pupils thought they had a better education compared with their siblings, nine (23.7%) considered it to be at the same level and two (5.3%) rated their education as poor in comparison. Thirty-one respondents (81.6%) thought they were treated differently at school compared with normally pigmented students in mainstream schools. In reply to an open question asking them to provide a description of this treatment, the responses were all positive ('We move to the chalkboard or it moves to us', 'We have CCTV, magnifiers and books with big letters to help us see as we do not see well', 'The nurse looks after our skin and tells us we mustn't play in the sun', 'The school here helps us' and 'I was bought to this school as I was beaten when I got things wrong in school at home'). Thirty-three pupils (86.8%) thought children with albinism at

primary level should be educated in special schools, while five (13.2%) said that it should be in ordinary schools but with particular attention. None chose the option 'in ordinary schools with no special attention'. Responses to the same question applied to secondary education were: 28 (73.7%), eight (21.1%) and two (5.3%), respectively.

Perceptions about albinism

Twenty-eight (73.7%) pupils said they did not know what caused albinism, eight (21.1%) correctly attributed their condition to a lack of pigment, two (5.3%) said it was 'in the family' and one (2.6%) thought the mother was short of something in the womb. Thirty (79.0%) pupils said their parents had never explained their condition to them. The others had looked at a book about the body and talked about albinism or had been told that albinism ran in the family. One said her mother told her she was a 'gift from God'. A few pupils mentioned a story about witchcraft ('Some people say it is caused by witch doctors. Some say that your parents have done something wrong to somebody') and the death myth ('They say we do not die. Is this true?').

In a multiple choice question, 31 (81.6%) pupils correctly thought the children of two parents with albinism would also have albinism (which is true only if both have the same type of albinism), six (15.8%) did not know and one (2.6%) thought the children may either have albinism or be normally pigmented. Answers to a similar question where only one parent had albinism were more varied: four (10.5%) thought the children would have albinism, four (10.5%) that they would be normally pigmented, four (10.5%) that they would have an intermediate (brown) skin colour, seven (18.4%) did not know and 19 (50.0%) were correct in thinking that some may have albinism and others not.

This group had a positive perception of the intelligence of people with albinism: seven (18.4%) believed they were as intelligent as normally pigmented people, 25 (65.8%) rated them more intelligent, with only one (2.6%) thinking they are less intelligent and five (13.2%)

saying they did not know. In terms of job opportunities, 22 (57.9%) thought people with albinism had the same opportunities as normally pigmented people, 10 (26.3%) that they were limited and six (15.8%) did not know.

Social and family interactions were more problematic. Thirty-three (86.8%) respondents thought people with albinism had fewer friends than others, and 14 (36.9%) found mixing at social gatherings such as dances and parties difficult. Although 15 (39.5%) pupils believed children with albinism were less loved by their parents and eight (21.1%) that they were sometimes ill treated by parents or family, eight (21.1%) thought they were treated the same as non-affected siblings and 14 (36.8%) felt they received extra care.

Perceived needs

In response to an open question about the most serious problems experienced by most people with albinism 27 (71.1%) mentioned poor eyesight, 24 (63.2%) skin problems due to sun exposure and four (10.5%) social problems (treated badly by parents, being shy, being subjected to name calling and feeling excluded in mainstream schools). When asked about the needs of people with albinism 28 (73.7%) felt medical assistance was required, 22 (57.9%) help with finding a job, 19 (50.0%) a forum or self-help group to provide support and 13 (34.2%) counselling about the inheritance of albinism.

Discussion

This study reports baseline information on the education, health, beliefs and perceived needs of a group of children boarding at a special school in northern South Africa, with a view to developing a model for successful intervention, particularly on sun protection, in an African cultural context.

Education: in special schools or mainstream?

The rural school where this study was conducted is a single disability school, catering for the needs of visually impaired pupils. The vast majority of

the children have albinism as this is the main cause of vision problems in this area. The school buildings are adapted to suit these pupils, with covered walkways and trees in the courtyard providing shade from the harsh tropical sun. Shutters on the windows helped reduce glare, as these children show extreme sensitivity to bright light. The school encourages and supports a positive attitude towards albinism, and enhances the pupil's self image and sense of self worth.

This group had an optimistic view of the intelligence of people with albinism, with most rating them more intelligent than others. Pupils were fully aware of the advantages of their education at this school, where having albinism was the norm, special adaptations were made to the classroom environment and teachers were aware of their visual impairment. Most rated special education, at least at primary level, as the best option for children with albinism.

This school seems to be one of only a few in South Africa where individualized educational programmes for children with albinism exist. Lund (Lund, 1997, 2001), in a study in Zimbabwe, concluded that these children can be successfully integrated into mainstream education, if teachers took into account their poor eyesight and sun-sensitive skins. Before subscribing to such an educational policy the support system needs to be scrutinized. In western countries adequate support can be taken for granted. Even in Zimbabwe with its well-trained teachers, it may be less of a problem. Most South African schools in the rural areas, however, are at the moment understaffed, crowded and have poor infra-structure, making it unlikely that such integration can be successfully implemented in the near future.

Sun protection

People with albinism are highly susceptible to unsightly and potentially disfiguring sun induced skin lesions and cancers. They have the skin type that always burns and never tans. The children in this study, living in a sunny, tropical area, have skin problems throughout the year and were fully aware of their extreme susceptibility to sun-induced

damage. Most took measures to reduce their sun exposure, applying creams daily, although half were aqueous creams with no SPF. Although pupils wore their hats regularly, encouraged by the teachers and nurse, many offered insufficient protection to the face and neck. Sunburn lines on the face clearly showed areas that were unprotected by the style of hat worn. The boys favoured baseball style caps which offer inadequate protection to the face and none to the neck. A demonstration of a Legionnaire-style cap with a long flap at the back, offering excellent protection to the neck, was greeted with laughter and derision, several adolescents declaring they would never wear such a hat. Peer image was clearly important; pupils are unlikely to wear protective clothing they consider ugly or conspicuous. Although their unusual appearance makes them immediately noticeable, the children desire to appear as normal and inconspicuous as possible. The children at this school do not have a uniform but wear their own clothes, including hats. At present the school is investigating the possibility of having a school hat, with a sensible wide brim, to be worn by all pupils.

Since application of a cream and wearing a hat were already part of their daily routine, the provision of suitable sunscreens and protective clothing should be effective in increasing protection and reducing the risk of skin cancer (Stern *et al.*, 1986). This is especially important during childhood as 50% of lifetime UV exposure is estimated to occur by age 20, making it desirable to focus prevention efforts on this group. There was evidence of peer support at this school, with pupils reminding each other to wear their hats, encouraging those with skin lesions to go to the nurse and even making sure those with skin sores applied their ointments regularly. The local hospital provides a generic SPF cream for those who find the cost of commercial preparations prohibitive. Although a 3-month supply can be obtained from the hospital, the cost and time involved in travelling to collect this deters many families. Regular visits by the genetic nurse to the school, bringing a supply of protective creams for the school nurse to distribute, would be an effective use of resources.

Visits by a specialist dermatologist would also help to treat potentially cancerous skin lesions early. Clinics in rural areas catering to a black population usually have no experience in the type of sun induced ailments suffered by those with pale skin, and cannot offer suitable treatment, often prescribing only an antiseptic ointment. There is also anecdotal evidence that both children and adults with albinism experience antagonism, avoidance and even rudeness when visiting local hospitals. If the suspicion that surrounds albinism extends to medical personnel then those with the condition will be discouraged from obtaining the treatment they require.

Eye care

The major disability for most people with albinism is their poor vision. Some of their many eye problems can be improved, for example by wearing tinted or dark glasses to help reduce glare and prescriptions to improve refractive errors. A separate study by the Department of Optometry of the local university aims to document the visual performance of these children and investigate ways of improving their vision.

Attitudes and beliefs about albinism

There are a number of myths and stories about albinism circulating in southern Africa. The superstition that a family with an affected baby has been cursed or is the victim of witchcraft is particularly strong in rural areas [(Lund, 1998) and this study]. The 'death myth', a belief that people with albinism do not die a natural death but simply go into the bush and disappear, is commonly heard in South Africa [(Kromberg, 1992) and this study]. These attitudes and beliefs need to be addressed directly when providing information on albinism in this region. Most pupils did not know what caused albinism, although their answers to questions about the children of parents with or without albinism showed that many were aware of the pattern of inheritance. This knowledge probably came from personal experience within their families and those of their peers. This baseline understanding provides a good grounding for enlarging their

genetic knowledge of albinism via talks, information sheets and discussion groups, using family case studies. An annual regional workshop on albinism and its management is planned and a group of university students with albinism are hoping to set up a self-help group, supported by the researchers on the 'Albinism in Southern Africa' project. Education of local communities about albinism should be part of this scheme, as it is amongst strangers that people with albinism often encounter antagonism and avoidance.

Health intervention in an African cultural context

The authors propose an interactionist approach (Babbie, 1995) to address the health and social issues of these children with albinism. Interaction focuses on small-scale intervention rather than on the society as a whole and base intervention on the meaning a person attaches to his/her condition. To be successful there must be compatibility between the objectives of these interventions and the values and beliefs about disease of the population for which they are designed, in this case Sotho children. African cultures often embrace supernatural rather than scientific or medical explanations for the onset of disease and are more concerned with cures than preventative measures. The children in this study are clearly well aware of the short-term consequences of sun exposure but may be less informed about the long-term risks of developing skin cancer. Given the fatalistic attitude within their culture to disease, an intervention strategy focusing on these future risks may not prove effective. An approach that stresses the immediate benefits of sun avoidance and protection may have more impact on behaviour. Short-term monitoring (diary recordings of sun exposure and subsequent episodes of painful sunburn or skin sores) and goals (reducing the number of burns, lesions or trips to the nurse for treatment of skin lesions) may be more appropriate. The children themselves must be convinced that staying out of the sun is a sensible and worthwhile choice. An intervention programme for these African children with albinism is thus likely to differ considerably

from one designed for European children, based on their different levels of knowledge and beliefs.

Given the strong and supportive group dynamics at this school, peer group discussion sessions leading to shared and agreed objectives on ways of managing their condition are likely to reinforce positive behaviour and lead to sustainable behavioural change. This will make the children active participants in the intervention, rather than passive listeners. Training in personal coping skills is also important if pupils in this relatively sheltered environment are to integrate into mainstream schools at secondary level and cope with adult life in the community.

A health-promoting school

This special school is an ideal setting for becoming a 'health-promoting school' [reviewed in (Lynagh *et al.*, 1997)] with the whole school focusing on sun protection by reinforcing and encouraging the good practice already present in the school. The school authorities could support its implementation by scheduling the timing of outdoor activities such as gardening and sport to the early morning or late afternoon, thus avoiding the time of peak UV exposure between 11 a.m. and 3 p.m. The provision of attractive indoor occupations for the pupils will encourage them to spend their leisure time indoors. The girls tend to spend their free time watching television, whereas the boys prefer to play soccer outside, leading to differences in the levels of UV exposure (P. M. Lund, unpublished results). The school and pupils could negotiate a policy on outdoor sports, e.g. 'No soccer without sunscreen'.

In their evaluation of a primary school sun safety programme in the UK, McWhirter *et al.* stress the role of teachers as effective communicators of the sun safety message (McWhirter *et al.*, 2000). Teachers at this special school are, however, all normally pigmented and do not suffer the same sun induced skin problems their pupils with albinism experience. Suitable information packs on albinism and its management aimed at teachers should form part of the intervention programme. They would, however, be transmitting information about sun protective measures without the need to do so

themselves and with no personal experience of sunburn. This information would carry more weight if presented by other members of the community with albinism and therefore first-hand knowledge of the health problems. A group of students with albinism at the local university are keen to give talks in schools. They would also be positive role models by showing that those with albinism can achieve university level, despite their visual disabilities.

The involvement of the parents is also crucial to a sustained health intervention plan, as the children often suffer skin damage when they return home during the holidays and are tempted outside to join their normally pigmented friends. If mothers can be educated and informed about albinism they will have more confidence to deal with their child's condition. Outreach programmes, support groups and workshops focusing on albinism and its management should be designed for an interactive, social learning process. These can become instrumental in creating an informed community willing to address albinism in an objective, scientific manner, but interpreted against the background of existing indigenous knowledge about albinism.

In conclusion, this special school provides an ideal environment to promote the health of children with albinism and implement a comprehensive, whole school approach to issues such as sun protection. There is a strong argument for re-orienting medical and welfare facilities so that personnel come to the school to deliver services in an efficient manner. Once an effective intervention programme has been established for this school, attention will be given on how to promote the programme in other settings. A further study is underway at a multi-disability school in the region, where physical, oral and visually impaired pupils are catered for. The intervention proposed here would need to be adapted to this different setting, where children with albinism are in a small minority.

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