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Living with Thalassaemia in Papua New Guinea, the experience of children, adolescents and their families

Running title: Thalassaemia in PNG

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ABSTRACT

Background

Thalassaemia, the commonest genetic blood disorder in Papua New Guinea (PNG) presents daunting challenges for the affected children, their parents and families, and the health system. We aimed to describe the quality of life of affected children and adolescents and the experience of and difficulties faced by their parents in the setting of a tertiary referral hospital in PNG.

Methods

A mixed-methods longitudinal study involving baseline questionnaire, then serial interviews with parents, children and adolescents living with β-thalassaemia attending Port Moresby General Hospital.

Findings

Twenty-one patients and their families were interviewed over a 6-month period. Most families originated outside the National Capital District and had migrated to be near the Port Moresby General Hospital and its blood bank services. Thirteen patients had at least one affected sibling and four families had experienced the death of at least one other affected child. No child was receiving chelating agents, and most had clinical evidence of iron overload. There were important impacts of thalassemia on quality of life, including very poor school attendance and some aspects of children's self-perception. Families faced significant burdens and made genuine sacrifices to care for their children.

Conclusion

Regular blood transfusions increase the life span of children with thalassaemia but there is a need to achieve a hyper-transfusion regimen coupled with chelation therapy. As for all chronic illness a focused and holistic approach is needed to improve the quality of life for affected children and their families.

Keywords

Thalassaemia, Papua New Guinea, Oceania, Quality of life

Key points

Thalassaemia occurs among children from malaria endemic regions in Papua New Guinea.

Children with thalassaemia share many needs with other children with chronic illness, including access to basic specialist services (blood transfusion and iron chelation therapy), and need for holistic care that promotes optimal growth, development, and quality of life.

Parents of children with thalassaemia in Papua New Guinea are often under financial and social stress to provide the best possible life for their children.

Introduction

Thalassaemia is one of the most common genetic disorders worldwide and presents significant public health and social challenges in areas where the incidence is high as it is in Papua New Guinea (PNG). There are more than 200 causative mutations in β-Thalassaemia alone.(1) Up to 7% of the world's population carry one or more of the gene mutations for thalassaemia (2) and 100,000 babies with severe forms of thalassaemia are born each year. In PNG it has been estimated that 68-80% of people originating in the north coast where malaria is endemic carry a thalassaemia gene mutation. A number of studies have indicated that the presence of a single or double α-globin gene deletion has a protective effect against malaria.(3)

β-thalassaemia is a chronic, debilitating and ultimately fatal illness which presents children and their families with major challenges. Globally it is estimated that only 12% of children born with transfusion-dependent β-thalassaemia have access to regular blood transfusion, while less than 40% of those transfused receive adequate iron-chelation therapy. More than 3000 adolescents and young adults die each year from iron overload, which affects cardiac, liver, endocrine and kidney function.(4) Impairment in neurocognitive development occurs, likely because of chronic anaemic hypoxia and fatigue, and other factors associated with chronic disease, and has been linked to the high number of hospitalisations occurring since early in life.(5)

This study describes a cohort of thalassaemia patients treated at Port Moresby General Hospital in the National Capital District of Papua New Guinea. We aimed to describe treatment received, to explore the perceptions of the patients' quality of life and to seek the views of their parents on the difficulties they face, their experiences and their understanding of their child's condition. There are no previous studies on children with thalassaemia, their experiences and quality of life, and few studies of children with chronic illness from PNG or Oceania.(6) We aimed to determine where improvements could be made in the care of children and adolescents with thalassaemia in PNG.

Methodology

A mixed-methods longitudinal study involving baseline questionnaire, then serial interviews at Port Moresby General Hospital (PMGH). PMGH serves a predominantly urban population of nearly 400,000 who originate from provinces throughout the country, and the hospital receives referrals from neighbouring provinces.

The first author (VN) conducted the interviews, mostly in Tok Pisin, with children and adolescents and their parents at regular intervals over a period of 6 months. An initial questionnaire documented demographic details, family history, clinical information and results of investigations. Interviews used both open-ended and direct questions (online Appendix). Certain questions were modified at the time of the interviews depending on the situation to ensure they were appropriate and not upsetting. The interviews took the form of empathic supportive discussions and parents and children were made aware that they could decline to answer any questions at any stage. Interviews were usually at times of treatment (such as receiving a blood transfusion), and care was taken that the clinical care the child required was not disrupted. If participants' responses were not clear, clarifying questions were asked at

the time or at a later interview. Interviews covered quality of life, participation in schooling, the parents' experience of living with thalassaemia, their understanding of the disease and their greatest concerns. Costs of care, particularly indirect costs such as bus fares and short-term accommodation in Port Moresby during an episode of care, were those reported by the parents. Responses from children, and the parents or guardians of the patients were recorded verbatim in writing and directly translated as needed.

We used a form of content analysis to best understand the issues; qualitative responses were grouped under common themes and reported with narrative description of expressed experiences, perceptions and verbatim responses. Quantitative data were entered into an Excel spread sheet and analysed descriptively.

Ethics approval

Verbal informed consent was gained from the parents and guardians of the children involved and special verbal consent were also gained from children aged 10 years or older. Ethical approval was obtained by the University of Papua New Guinea School of Medicine and Health Sciences Research Committee.

Results

Over 6 months, 21 children with thalassaemia and their families attending the hospital for acute or chronic care were interviewed several times. A comprehensive summary of the questions and responses is in an on-line appendix, and key issues are highlighted below.

The patients' median age was 7.7 years (interquartile range of 4.7 - 14.4 years). Seven children were 5 years or under, 7 were aged 6-10 years, 2 were aged 11-15 years, and 5 patients were 16 years or older. 14 were female. Nine were from Central Province, and eight from Gulf Province. Many families had migrated to Port Moresby so their children could receive regular blood transfusions.

Thirteen children had a family history of thalassaemia, and eight had affected siblings, including four families whose other children had died from thalassemia. No advice about recurrence risks had been given to any of the 12 mothers who delivered in health facilities. During the 6 months of the interviews two mothers became pregnant, two new cases were diagnosed, and one of the 21 children died.

Eleven children had been diagnosed on clinical grounds and with full blood examination, eight had haemoglobin electrophoresis confirmation, and one had DNA confirmation (in the Philippines).

Clinical manifestation of thalassemia and management

All patients complained of lethargy or tiredness. Frontal bossing and maxillary hypertrophy were present in 14, hepatomegaly in 12 and splenomegaly in 13. Twelve children were failing to thrive, eight had jaundice and four had bleeding episodes. All 21 children were receiving blood transfusions, and none had received any iron chelation therapy.

Social interaction and schooling

Twenty children were not currently attending school. Only one was attending primary school, and in grade four. Five children had attended elementary school (kindergarten) and four had attended prep grade but proceeded no further, and five had never been to school. The need for recurrent blood transfusions interfered with continuation of education. Parents also said they were worried that

accidents may happen at school. Eighty per cent (16/21) said their child was too weak to walk to school and most cited the child's distended abdomen as an indication of that general weakness. Some children refused to go to school despite their parents' encouragement and four said they quit school because they could not concentrate in class. Another five said they were stigmatised and called names at school.

> *"Em lusim skul bikos ol classmates blon em save kolim em "Bel Mama".. A taim blut sot em bai weak na slip planti na no nap wokabaut or bai kisim skin hot na sik so mipla sa kisim em kam long hausik..em misim planti klas tu."*
>
> *She left school because classmates called her "Pregnant Mother" When her blood is low she sleeps a lot and can't walk and easily gets sick and feverish. So I keep her at home and she misses plenty of school."*
>
> *'Why em no go school? Bcoz of sik so nogut "by mistake ol bai bamim em na spleen bai bruk na em dai."*
>
> "Why does he not go to school? Becasue of his illness - so it's not good if by mistake he gets bumped and his spleen bursts and he dies."
>
> But there were also good experiences of school and acceptance within the local community:
>
> *"Long hausik na autside ol save lukluk strong..but long ples he is accepted na ol lain lo ples save olsem em sick mangi..tupela wantaim even lo skul too..tisa na students save understand and pilim sori long tupela. Ol save was long ol tu...tupela no save pay busfare."*
>
> "When we are outside from the village or at the hospital the people stare at him and us but when we are at the village, he is accepted well and the teachers, students and the community are aware of his older brothers who had similar sicknesses and died therefore they are sympathetic and are empathic that's why the PMV (public bus) rides for him is usually free when we come for blood transfusions."

Self-perception about the illness

The children were asked what they perceive about their illness. Only one child saw thalassemia as no obstruction to his social, physical, education and future personal development. He dreamt of becoming a doctor therefore he attended school daily and took part in all activities. The other patients saw themselves as restricted due to the illness and were mostly confined to their homes. Despite this considerable reduction in normal activities, they did not see themselves as being a burden to their family.

Family's financial situation

Seven of the families supported their children by having regular jobs and earning a fortnightly salary. Another seven had informal ways of making money including gold panning, operating a road-side trade store, lending money, and selling beetle-nut. An additional seven families survived as subsistence farmers.

The families that had migrated to the National Capital District had reduced the total cost of return transport for blood transfusion to the median of K5.50. For the three families living outside Port Moresby the indirect cost of blood transfusion and medical care was up to K400 per month. Six other families spent K100-150 for each episode of care.

Knowledge about thalassaemia of prognosis

When the parents were asked if they knew their child's diagnosis, nine said that they needed more explanations, seven understood about thalassaemia, and two refused to accept that their child had the condition.

> *"It's inherited and is passed from generations...tumbuna, tumbuna..There are 3 types. Group A na B, endemic...no proper/specific diagnosis and classification so mipla no save which type em gat...." (Idont know which type hs has)*
>
> *"Em igat thalassaemia-nogat treatment-only blood..."*
>
> *"He has thalassaemia. There is no treatment - only blood".*

The parents were asked if any health workers had explained the prognosis of thalassaemia to them since their child was diagnosed, and if they were aware of the limited life their child would have, even with regular blood transfusions. Fourteen of the parents reported having been informed whilst eight said they never knew the prognosis and were sad and crying when this was discussed. Two parents were not willing to accept that their child's long-term prognosis was poor, or that the condition was life-limiting.

Many parents commented that thalassaemia does not get the support from the government or doctors like other diseases.

> *"Other sick like TB, HIV, Cancer, sik bun nating ol doctor na nurse I save busy long ol, mipela nogat true...sampla taim sapos mipela no lukim ol save pes mipela save wait longpela taim stret. Dr Laki tasol save helpim mipela hariap tru taim em stap lo wok."*
>
> "The doctors and nurses are busy with other sicknesses like TB, HIV, cancer and malnutrition but not with us. Sometimes if we don't see anyone who recognises us we have to wait for a long time. When one of the doctors who knows us is working he helps us quickly."

Family coping strategies

Families had different strategies to cope with the fact that their child or children have thalassemia and a poor prognosis. Twenty parents expressed that although unfortunate, they will do all their best to give their child or grandchild a good lovable life and try to improve and prolong their child's life if possible. Nine parents requested for medical reports to seek financial assistance from their local member of parliament, the provincial administrators, companies or financially capable relatives to consider further investigations and treatment overseas. Some parents discussed their interest in forming a thalassaemia foundation in PNG so they can affiliate to other thalassaemia foundations from around the world for continuation of research and access other treatments for thalassaemia. Two said God had a reason for thalassemia to occur within their family and only He will solve the issue as He see fits.

Some parents had consulted traditional healers:

> *"Thalassemia em olsem blut sot..spleen em bikpla olsem na em kilim blut...so bai yumi go kam long kisim blut. Spleen sa drainim blut blo ol so mipla papamama think its spleen so mipla kisim pikinin go long relatives, pay money na ol rubbim spleen...but nogat change..money waste nothing."*

> "Thalassaemia meant that blood is low due to the enlarged spleen which is destroying the blood cells therefore we are frequently receiving blood transfusions. The spleen drains the blood therefore we the parent thought it was spleen so we took the child to known persons or healers here money is paid and the spleen was rubbed with no change. We wasted a lot of money on the healers or heal our son."

Seventeen parent couples were still married and living together. One of the parents in the other families had died. There were no divorces or separation among families in this study.

Parents' major concerns

The main concern among all the parents was the prognosis of thalassaemia that will result in their child's death, and the unavailability of the iron chelating drugs.

> *"The haematologist promised the patients iron chelators to bring to PNG but never did and while waiting we lost 2 of our children who were monitored by the doctor and were receiving blood transfusions."*

Eighteen parents were worried that their children may get infected with blood-borne infection. They also worried that one day they may be not enough blood supply to transfuse their children and that they have to pay people to donate blood for their child, thus increasing the financial burden to their family. Fifteen parents were concerned about their child's physical appearance, this was often expressed as worry relating to the physical deterioration of their child in front of their eyes, with the implication that their child was slowing down and reaching the end of his or her life.

Most parents were not primarily concerned about their child's absence from school, separation from their families because of seeking treatment in Port Moresby, or separation from their friends.

Positivity about illness and services

Despite the chronicity of thalassemia with all the physical symptoms and difficulties the children and families faced, parents remained positive about certain issues. Nineteen stated their appreciation that the health service is free with free blood transfusions and blood tests. Fifteen said they received better attention and treatment from concerned regular staff. Parents also expressed concerns for others: several said their familiarity with the hospital enabled them to assist other patients who looked lost. The majority of the children stated they knew people and have friends in the hospital and eight excitedly explained that their parents buy them special food when they are in the hospital, allow them to hold and use their phones to play games or watch movies and when in hospital their family are always around with them.

Discussion

Thalassaemia presents major challenges for the affected children and their families in PNG and similar countries in Oceania. Access to blood transfusion is vital and many families migrate from their villages to near the major hospitals for blood transfusion. The three standards of care for thalassaemia are (i) to maintain a hyper-transfusion state, keeping the haemoglobin above 10g/dL, (ii) chelating agents to prevent haemosiderosis, and (iii) holistic care in a multidisciplinary way.(7, 8)

Iron chelation is expensive or difficult to administer. Deferoxamine is the cheapest chelating agent available but has to be given subcutaneously for 5-7 days every week at a cost of around US$150 per week for a 20kg child. Oral chelating drugs (Deferiprone and Deferasirox) are available but at a prohibitive cost of around US$300 per week. The average annual per capita income in PNG is estimated by United Nations Development Program (UNDP) of US$2,386 (K8422, or K162 per week).(9) This also provides perspective on the current cost of an episode of care (typically required monthly), which for families residing outside Port Moresby is at indirect costs similar to or exceeding the average weekly earnings.

Our study illustrates the major effects of thalassemia on quality of life for affected children and their families. Only one of our patients was attending school and chronic fatigue was a major feature. Fear of stigma and of unintended serious injury featured in the parents' concerns. Our findings are similar to a study of mothers caring for children with thalassaemia in Thailand which identified lack of knowledge, psychosocial problems, concerns for the future, social support systems, financial difficulty and concerns on the effectiveness of health care services.(10)

As in many low- and middle-income countries, the focus for child health services has been on infectious diseases and malnutrition. Yet there are many children with chronic illnesses like thalassaemia who require long term, holistic and readily accessible care. Paediatricians have an important role in coordinating care and advocating for better services for such children.(11) Most reforms in integrated care for chronic non-communicable diseases have focused appropriately on adult diseases.(12) In addition there has been a welcome global focus on neglected tropical (infectious) diseases, all of which are chronic and debilitating. Unfortunately, chronic non-communicable diseases affecting children and adolescents have received little attention, and hospitals and health services consequently have been slower at developing programs for children with epilepsy, developmental disability, sickle cell anaemia, thalassaemia, diabetes, cancer and other chronic conditions. The era of the Sustainable Development Goals, the re-design of the World Health Organization's Child Health Program,(13) and new momentum from families and affected people in many countries provide opportunities to address this.

The realisation that a child has thalassaemia is devastating for families. Having a second and sometimes a third affected child must compound a family's stress and predicament. Some parents suggested the establishment of a thalassaemia foundation or local support group which could link into the International Thalassaemia Federation.(14) They also suggested a separate thalassaemia clinic with designated staff who are familiar with the families and the disease. Educational materials could be made readily available and opportunities provided for holistic, multi-disciplinary care, including genetic counselling and family planning advice.

Despite the difficulties in their lives, most of the children remained positive and looked forward to their hospital visits for blood transfusion. They have friends in the hospital and saw their visits as an opportunity for them to eat special food and or have some fun activities. Non-communicable diseases

are increasingly common in children requiring a special focus, and should be addressed in a humane and holistic way within the available resources in PNG and other countries in Oceania.

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