Grace's Social Determinants of Sickle Cell: A Pen Portrait

In one person, sickle cell disorders (SCD) can present as chronic illness, in another a disability and for some people it is an acute and serious life-threatening illness. All three can also be present in the same person but social and environmental conditions are important to understand, as no individual is the same. To illustrate how SCD is affected by the biopsychosocial as well as socio-cultural and environmental conditions according to the social determinants of health of sickle cell model, the hypothetical pen portrait of six-year-old Grace Lekan who lives with her parents is given.

Individual factors

Grace was born in a sub-Saharan African country where they had a new-born screening scheme for sickle cell disease. However, very soon after her birth her family fled to a neighbouring country due to the internal armed conflict, so the health service couldn't contact her parents to inform her she had sickle cell disease having inherited one sickle gene from each parent. In Grace's first few months of life she was a thriving happy and healthy baby, giving much joy to her parents. At around 6 months of age this all changed when her hands and feet began to swell and she would cry uncontrollably. The family began to search for a diagnosis for their daughter's symptoms, being told different, conflicting things by neighbours and health professionals.

Biological factors

Grace also started to experience several infections and temperatures and a local health clinic initially diagnosed her with malaria. With repeated episodes of infection and pain, everyone but her mother saw her as a difficult, fussy child and people criticised the way her mother brought her up. At three years old, Grace's stomach began to swell and she became almost unresponsive so was rushed to a hospital where she was tested. Her mother was told Grace had sickle cell disease (HbSS) and needed particular care. Mrs. Lekan had other children and this is her first experience of caring for a child with sickle cell disease. While Mrs. Lekan was a carrier for the sickle cell gene, no one in her extended family had ever had the disease, so she did not know how to care for someone with the disease. The doctor explained to Grace's parents that each parent was a carrier. Mr. Lekan felt fine and did not believe the doctor's scientific explanation that he carried a disease. He concluded that he could not have produced an ill child, so accused Grace's mother of infidelity.

Social factors

Grace's mother tried to find out more about SCD. With almost 90% of children born with SCD in sub-Saharan Africa not living to their 5th birthday, she found it hard to find anyone with a positive prognosis for Grace. Her community had all kinds of explanations for why Grace had the disease, ranging from her being cursed, to her mother having eaten the wrong food during pregnancy and she was told people with sickle cell are spirits that come and go. Doctors would not say Grace would live beyond 5 years old either, but did say that those with more access to hospital care and a healthy diet and clean house would live longer but would possibly experience strokes and organ damage as they grow up. Despite the fatalistic ideas of everyone around her, Mrs. Lekan found out that making sure Grace was well fed, drank plenty of fluids, didn't get too hot or cold and kept happy was something she could do for Grace even without money for medical care. Mr. Lekan tried to do his best to take care of his family by taking on extra shifts as a taxi-driver but he could not always buy

the medicines that Grace needed. Ashamed to admit it, he once asked Mrs. Lekan if it was not better that Grace died instead of living.

Psychological factors

Whilst many people, including Grace's dad, had given up on Grace, her mum gave her extra special treatment, always making sure her hands were warm, going without food so Grace could eat, and trying to make her enjoy something every day. Neither the doctors or community could actually predict how Grace would fare with SCD. Each individual reacts differently, some people with HbSS, the more extreme type, can go on for years without being affected and some with what can be considered a less severe type, such as HbSC, can have continual problems. Even some carriers of the sickle cell gene can experience some symptoms, Grace's mother used to feel pain in her joints when the rain came. She didn't complain but used that to remind herself that Grace would need extra layers in that moment. Grace lived happily for the next 3 years without experiencing another SCD related complication and the household became peaceful. As they saw Grace thrive, Mr. Lekan apologised to Mrs. Lekan. He thanked her for giving him a beautiful and healthy little girl and he looked after Grace and the other children in the evenings to give Mrs. Lekan a break.

Individual lifestyle factors

As Grace grew older, she began to reject the special treatment her mother was giving her. She would throw off the extra layers of clothing her mum put on her when she went off to school, even if that made her joints go stiff on cold mornings. Grace was usually a lively child but was not allowed to play outside whilst at school or at home so she did not make many friends. Her teachers do not understand why she seems distracted and lethargic in class, and is always having toilet and water breaks. Her mother told them that the doctors say that Grace needs to drink water and she offered to talk to them about sickle cell. As a refugee, Grace is typically ignored by the teachers and the school authorities, and they are also not used to having a student with SCD because they know most of them don't reach school age. Because her attendance was poor and she is sleepy during class, her teachers do not allow her to pass her year, and she has to stay in the same class.

Social and community factors

When the family fled from war to a neighbouring country, they had to move around to find someone to stay with. When Grace was 6 years old, they were staying with a family contact but in return for shelter Mrs. Lekan and Grace were doing the cooking and cleaning in the house. As Grace grew older they expected her to take on more of the housework. Whilst washing the dishes one day the cold water triggered a crisis in Grace's wrists and Grace needed emergency medical attention. The family did not know where to get help so quickly, there was no bike transport and they did not have anyone who could lend them money to pay for the fees. Unfortunately, Grace died suddenly. Mrs. Lekan was eyed with suspicion by her husband's extended family and they accused her of witchcraft for having a child with the 'strange bone disease', causing big arguments with her husband. Devasted by Grace's death, Mr. Lekan wanted to find another wife who could give him 'normal' children so he left Mrs. Lekan.

Living and working conditions

While Mrs. and Mr. Lekan followed the doctor's advice and had even bought Grace a net to sleep under to prevent malaria, they did not know everything about SCD. One of the things that also contributed to Grace's final crisis was that she had had several minor sickle crises leading up to that

after a gastrointestinal infection she caught because they had poor hand washing facilities The vomiting caused her to become severely dehydrated which caused her cells to sickle. She did not need hospital treatment at the time, but she had not fully recovered by the time the cold water caused the major crisis. Despite having a net, she shared a room with her siblings and parents. The overcrowded living conditions with paraffin burners had caused her repeated respiratory infections. One of the complications of her last crisis was acute chest syndrome which was made worse by the existing bronchitis from the previous infections.

National and global politics

Grace's living to the age of 6 years was beyond the expectations of her community and health professionals but her death, and those of her peers with the disease, did not cause any waves of outrage nor did it contribute to any national or international statistics on the condition. They are not taken into consideration during a pandemic either and there is no international outcry or concern about this population of people. Whilst there is a significant global awareness about child mortality in sub-Saharan Africa the contribution of SCDs to these statistics, despite being more than HIV/AIDS is not highlighted. Preventing the deaths of children like Grace is not on many people's agendas and this looks to continue. This is the same in the country where Grace lived and all over Africa, the Middle East, India and the Americas where the condition is more common.

SCDs have not been prioritised in global public health research, policy and resources and few questions this neglect. Even in high-income countries, because SCDs mainly affects people of African origin, remains a marginalised condition with limited support and funding for patient support groups or the voluntary sector. This has led to a lack of awareness about how people, families and communities are affected and the need for more holistic support. The global health focus for Sub-Saharan African countries is set by international funders with priorities like HIV and child health issues like malaria, reinforcing medical neglect and community stigma of the condition. While some basic treatment is accessible, specialised treatment is limited and unfathomable to most ordinary people. Global, regional and national inequalities lead to people experiencing extremes of poverty and then more illness, disability and deaths as a result. The 'burden' of SCDs is one of neglect and, as illustrated, undertaken entirely by the individual with the condition, family members and community institutions in the absence of holistic global health.