Addressing adherence to treatment: a longstanding concern. 
The health care professional

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Abstract

Today a patient born with thalassaemia major can expect to have a near normal life expectancy and remain free of complications of iron overload with good monitoring and excellent transfusion and chelation regimes. Unfortunately patients still develop complications as a consequence of iron overload including endocrinopathies and cardiac failure. The main reason behind this failure of effective treatment is inadequate treatment. This can be due to either clinician related factors, patient related factors or lack of adequate provision of medicines and services. In this short paper I will highlight where the challenges lie with regards adherence to treatment and suggest approaches to manage this.

Introduction

Effective management of thalassaemia syndromes has converted a disorder that was fatal in the 1950s to that of a chronic long-term condition associated with good survival. Instrumental to this outcome has been the development of iron chelation regimes and monitoring tests that have allowed clinicians to adjust chelation therapy and where possible prevent the onset of complications.

However, despite these advances that now allow patients with thalassaemia syndromes a near normal life expectancy with minimal or no comorbidities, we still see patients develop new endocrinopathies, myocardial iron loading and indeed die as a consequence of iron overload. A number of factors contribute to the poor outcome in some patients living in resource-rich countries where there is good access to care and medicines. These factors can be defined as either clinician related factors or patient related factors.

Clinician related factors that result in a poor outcome are multifactorial and include:
1. Inadequate understanding of thalassaemia and its treatment
2. Insufficient experience with the chelation regimes and management of side effects leading to inadequate chelation.
3. Ineffective use of and interpretation of monitoring tests such as ferritin trends, liver function, renal function, endocrine function
4. Lack of availability of MRI monitoring tools such as R2, T2 for liver and cardiac iron
5. Ineffective communication with the patients or the family around what the tests mean for the patient as an individual.
6. Inadequate communication with patients around what the factors are that affect the patients ability to adhere to treatment

Patient related factors are more complex and even well informed and normally compliant patients can have episodes of poor compliance. Factors that affect a person’s ability to adhere to treatment include:
1. Misconceptions of thalassaemia and its treatment and life expectancy (illness perception)
2. Misconceptions with regards chelation drugs, how they work and how to manage side effects
3. Lack of self efficacy and a feeling of helplessness
4. Family dynamics
5. Peer pressure
6. Stigma related to having a disorder
7. Work and lifestyle
8. Depression and anxiety

Discussion

It is therefore critical that in an era where excellent clinical outcomes are possible that both clinician and patient related factors are tackled to help improve patient outcomes.

Tackling clinician related factors

The UK Thalassaemia Registry first highlighted poor outcomes in patients with thalassaemia due to a lack of specialist expertise. Clinician related factors in the United Kingdom are being tackled by ensuring clinical networks are developed and all patients with thalassaemia are supervised by a specialist centre even if they are transfused at a local centre nearer to home. This means that lack of specialist knowledge in thalassaemia, its treatment and monitoring becomes minimal. The development of specialist teams will ensure that patients have access to clinicians that are knowledgeable about their condition and can advise the local centre teams in order to optimise clinical outcomes. Specialist centres should have access to the full range of monitoring tests including MRI monitoring tools. In addition the establishment of the National Haemoglobinopathy Registry (NHR) in the United Kingdom and the need to report serious adverse
events as part of a key performance indicator will ensure that learning happens nationally into why some patients have poor outcomes.

Communication issues between patients and clinicians are more difficult to rectify but the hope is that specialist teams where there is good psychology support will help to improve communication around factors that affect adherence. Improving communication with individual patients takes time and it is critical that a good understanding of the patient’s perspective of their disorder is gained in order to improve adherence. The role of clinical psychologists that are integral parts of the team is critical to helping ensure this. From a clinician’s perspective to listening to and educating the patient to clarify any misconceptions is critical to helping improve both self-efficacy and illness perceptions.

**Tackling patient related factors**

Many factors will impact on a patient’s ability to adhere to the treatment of a complex and often difficult and stigmatising health condition. One of the key interventions starts even before a child with thalassaemia understands they have the condition. The role of parental education and confidence building is critical in helping a child have a pragmatic and positive approach to their disorder. Where the parents view the disorder as a stigma and are either overprotective or disengaged with treatment or clinical outcomes the child will be negatively influenced with regards their approach to their condition. Good management starts from the time of diagnosis of a patient with parental education and support. All areas of thalassaemia care should be tackled; from transfusion regimes, iron chelation and side effects and challenges with this, how to encourage the child to be like other non-thalassaemic siblings and how to ensure that transfusion episodes and chelation treatment are treated with the minimal amount of stress.

As the child grows and becomes more aware of their condition, parents, doctors, nurses, play specialists and clinical psychologists should all be involved in helping tackle any issues that affect the patients perception of their disorder. Patient education on their disorder and expectations on life expectancy and quality of life are critical in improving health beliefs and converting illness perceptions to wellness perceptions. Part of the education over the childhood years is the critical role of iron chelation therapy in helping keep the patient well.

Good engagement and communication with the patients and families will help to clear up misconceptions and stigma relating to thalassaemia and the need for treatment. Patients should, by the time they reach teenage years, be taking more responsibility for their treatment and if on oral medication should be self-medicating at the latest by the age of 16 years.

Peer pressure is a difficult issue for all young adults and regardless of the presence of a long-term condition is a challenge for all patients. Many young adults when they attend schools, university or colleges will not tell colleagues that they have a health condition and this can often result in a failure to comply to treatment. Good patient education from a young age and psychological support if needed can help to reduce the impact of negative peer pressure on patients. Patients who are treated in large thalassaemia centres where there are patients of their own age and older role models are often subjected to positive peer pressure when they see their peers or older thalassaemia being successful.

Adult patients may find adherence to treatment challenging due to work or lifestyle issues. Appropriate adjustments should be made in treatments to fit in with the patient’s lifestyle. Detailed discussion should happen to look at when chelation can be taken either during the working day or at home. Often it will mean mapping with a patient what happens during their average week.

Compliant patients may develop episodes of non-compliance either periodically or for a longer period of time and these episodes are often related to anxiety, depression or serious life events. During these periods the role of clinical psychologists in helping the patient manage the complexities of treatment and life events simultaneously is critical. Nurse specialists who know the patients well and can help provide support and encouragement as well as doctors who can help to make treatment less onerous will help the patient to return to a more compliant state. Key factors that will help improve patient adherence to treatment and ultimately clinical outcomes are initiatives such as the expert patient programmes.

**Conclusions**

The management of adherence to the complex and rigorous aspects of treatment needed by thalassaemia patients is best managed with a multidisciplinary team that consists of the patient, their family and friends, doctors, nurses, counsellors, and clinical psychologists. At the heart of this strategy is education of patients and their families so they become experts in their health and disease prevention strategy.

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