Cyflwynwyd yr ymateb i ymgynghoriad y Pwyllgor Iechyd a Gofal Cymdeithasol ar Cefnogi pobl sydd â chyflyrau cronig

This response was submitted to the Health and Social Care Committee consultation on supporting people with chronic conditions.

CC48: Ymateb gan: | Response from:

Hereditary Anaemia Service for Wales working under Cardiff and Vale Health Board.

By Email

9th January 2020

Len Richards
Chief Executive
Cardiff and Vale University Health Board

Dear Mr Richards

Health Services Caring for People with Haemoglobin Disorders - Cardiff and Vale University Health Board

Enclosed is the final report of the review of services for people with haemoglobin disorders which took place on 1st October 2019. We hope that this report will provide assurance about aspects of the services which are working well, and will be helpful in highlighting improvements you might wish to consider.

The report will be made publicly available by the Quality Review Service (QRS) through their website in March 2020. We recommend that you place the report in the public domain, and deal with any associated publicity, prior to the report being placed on the QRS website.

The report reflects the findings of the review team, all of whom were invited to comment on a first draft. Trust comments on the draft report have been taken into account and the report has been considered by the UK Forum on Haemoglobin Disorders Peer Review Steering Group. The Trust may, however, appeal if you consider the report is unreasonable or disproportionate in the light of the available evidence. Appeals must be submitted within four weeks of receipt of this letter, and will be considered by a sub-group of the QRS Board. Appeals should be addressed to Charlotte Henderson, Business Manager, QRS at charlotte.henderson2@nhs.net and should include a statement of the reasons why you consider the conclusions reached were unreasonable or disproportionate.

The issues identified in this report are the responsibility of the Trust working with its commissioners. There will be no follow-up by the UK Forum on Haemoglobin Disorders or the Quality Review Service, although we would be very interested to hear of progress you are making.

Please will you convey our thanks to your staff for their help with all aspects of the quality review programme. If you have any queries about this report or any action you are now expected to take, please do not hesitate to get in touch.

Yours sincerely

Subarna Chakravorty

Dr Subarna Chakravorty, Dr Emma Drasar, Dr Rachel Kesse-Adu and Dr Mark Velangi
Clinical Leads for Haemoglobin Disorders Peer Review Programme

cc: Trust Leads

To the Health and social care committee holding an inquiry into supporting people with chromic conditions.

This evidence concerns the care of people with haemoglobinopathies both in the community and in the hospital environment.

We welcome the inquiry and wish to show evidence of a positive change in the care of children and adults with these conditions in Wales, however considerable problems persist.

Why is access to commissioned specialist care in hospital and the community important for people with haemoglobinopathies?

Haemoglobinopathies are the commonest serious genetic disease on the planet. They are the commonest cause of serious genetic disease in the UK. They affect all racial groups but are more common in people of BAME ethnicity. Over the last 20 years there has been a steady increase in cases of haemoglobinopathy in Wales. There are children and adults with haemoglobinopathies all over the Principality, both North and South and all need ongoing care of their complex multisystem disorders.

Haemoglobinopathies are a group of disorders in which haemoglobin, the oxygen carrying molecule in blood is either not made (thalassaemia) or made differently (sickle cell) in such a way that causes disease. Any organ in the body that has a blood supply can be damaged in the haemoglobinopathies (ie all of them). The most common disorder is sickle cell disease. Furthermore, the patient group that experiences these diseases are from ethnic minorities and experience a range of disadvantages

The treatment options for these patients are limited. Research into finding therapies has been poor over the last 50 years. Patients are from severely disadvantaged communities in developed countries while the majority of patients globally live in low/middle income countries with poor resources. Although they represent a serious burden of disease in Europe and the USA as well as in India, Brazil, Nigeria and a number of equatorial countries the pharmaceutical industry has not been interested in finding treatments.

Fortunately, there has been a change in this situation over the last 10 years with a large number of multicentre global trials beginning to bear fruit and new drugs and therapies beginning to be licenced.

Nevertheless, at the moment the mainstay of therapy remains transfusion of blood and the use of hydroxycarbamide. These treatments have side effects – regular clinic attendance, need for monitoring and iron overload. None of these therapies is curative and the life expectancy of sickle cell and thalassaemia patients is reduced.

Commissioned Services in Wales

Wales has initiated national screening for haemoglobinopathies. All pregnant women are risk assessed and screened for carrying the disorder and all new-born babies are screened at day 5 of life by the heel prick blood spot test.

There are about 80 children and around 100 adults with haemoglobinopathy in Wales.

Over the last 5 years clinicians in the north and south of Wales have collaborated with the Welsh Health Speciality Commissioning Committee to set up a commissioned service within the country. Clinical services integrate into a larger multidisciplinary team for advice and guidance. North Wales

to the Manchester HCC and south Wales to the west London HCC. The service has been peer reviewed (see attached PDF files). These highlight the shortfall in services. Services in England have undergone significant change since 2018. However the All Party Parliamentary Group published the report of “Nobody is listening” (https://www.sicklecellsociety.org/no-ones-listening/) in 2021 following the tragic death of a number of patients with sickle cell disease in English hospitals shows that further work is needed.

Current Problems

There still significant issues with the service in Wales. These are:

Community service

There are no community services in adults to allow treatment of patients nearer to home. Data from other countries, particularly Jamaica, show that early treatment of sickle cell crisis in a day care setting offers better control and less disruption to work, education and social interaction. Patients with thalassaemia, who depend on regular blood transfusion for survival, need to attend hospital every few weeks. Although we have set up a clinic to allow young patients to attend on Saturday, rather than a work or education day, this still disrupts life significantly.

There is a paediatric community service.

Social Care

A large percentage of the children are homeless. This is often due to their status as newly arrived into the UK. There is a large amount of work for the team to navigate the social care and housing systems.

Psychology service

The psychological impact of the chronic pain, with acute exacerbations, associated with sickle cell disease cannot be overstated. Currently, there is no psychological support for these patients although this is in set up under funding from WHSSC but the post cannot be appointed to as there have been no applicants.

MRI iron scans of heart and liver for children, and of the heart in adults are not available in Wales. Children have to go to Birmingham and adults have to go to London to have annual scans to look for iron loading.

Acute care

Admission at times of crisis for the Adult patients is difficult. The emergency departments and medical admission wards are under immense pressure and staff there are usually lacking in deep knowledge about the conditions.

There is no acute pain team in paediatrics in Cardiff for sickle cell.

Education of health care workers and the general population is essential to address these life inequalities. Institutional racism remains a stain on the service and we need to reach into schools and universities to ensure affected individuals are appropriately supported.

Key Recommendations:

1 – action the WHSSC funded psychology service and recruit (CAV to do)

2 – pain control service for paediatric sickle cell patients

3 – Set up T2* MRI scan/FerriScan liver and heart iron measurement in Wales for children and adults

4– commission a community service for adults

5– social work help for homeless families with children with haemoglobinopathies

6- Action to alleviate the acute medical admission crisis in Wales

7 – Education to address institutional racism that these patients experience

Dr Philip Connor – Consultant Paediatric Haematologist, Children's Hospital for Wales

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