Bringing life-long, joined-up care to people with rare and complex health conditions

Ensuring integration delivers for people in need of specialised services

Introduction

The Specialised Healthcare Alliance is a coalition of charities and corporate supporters campaigning on behalf of people with rare and complex conditions in need of specialised care. These conditions often have no cure – they are for life – and symptoms may get progressively worse over time. People living with such conditions require a complex combination of physical and mental health care, as well as community and social care services. Their care must be ongoing, and their changing needs must be assessed regularly.

Yet, within the present system, care is disjointed, and patients can face great difficulties in accessing the support they need. Our members report that some patients can be discharged from specialist clinics and left in the hands of more generalist services in primary or community care settings, without the experience of managing someone with their condition.

The SHCA believes that the moves towards integration of health and care services, including those proposed in the recent NHS White Paper, represent an opportunity to join up specialised and less specialised services in a way that will help to deliver equal access to high quality, patient-centred care that improves the quality of life – and therefore overall health – of people with rare and complex conditions.

However, any such steps must be carefully planned, to ensure integration delivers meaningful benefits in ways which preserve the specialised nature of the health and care services that need to be delivered.

This briefing is intended to help shine a light on the complexity of these issues. It sets out:

- A number of case studies – informed by our members – which aim to demonstrate where integration can deliver improvements in outcomes for people in need of specialised health and care services
- A range of recommendations, based on our members' insights

Case studies

Case study 1: Brittle bone disease

Care requirements: orthopaedics, physiotherapy, dental care, mental health, social care

Brittle bone disease is a genetic disorder that leads to fragile bones that break easily. The disease manifests in different ways between patients. Some patients experience physical disability requiring walking frames or wheelchairs from a young age. The Brittle Bone Society report that many patients receive manual wheelchairs from the NHS, but this leaves them trapped indoors, requiring carer support in order to leave the house, with severe implications for the patients' mental health. The charity raises money to fund specialised electric wheelchairs that help allow affected patients to lead a near normal life. One patient said:

"Simply being able to go out and have a coffee [that was not possible before] has now become something I can do without much effort, not only helping with my overall independence and mobility but also with my mental wellbeing. Before this I have often faced the overwhelming feeling of being trapped simply because I could not leave the house

without help. This in itself can be debilitating, as you wake up every day realising you are stuck in a loop. Having this one extra tool in my arsenal has allowed so many other parts of my life to open up. I am 33 and now I am able to do more, thanks to my new equipment, I can finally feel like I am progressing.

Investing in available technology that improves patients' quality of life – such as a specialised wheelchair – can therefore reduce the need for community and social care support, as well as having a significant and positive impact on mental health.

Another symptom of brittle bone disease is brittle teeth. The loss of teeth can have implications for a patient's diet, and their overall physical health and mental health. However, patients are frequently not referred to a specialist dentist. One patient said: *"The local dental hospital was able to extract teeth but this would mean leaving me with no bottom teeth to bite with. When I asked about being referred to a specialised dentist I was told I'd have to contact them all to see they would accept me with my condition. I don't seem to be able to get any referral and no one is willing to take me on."*

Another patient said: *"I've lost five teeth and only a few days ago another front tooth cracked which has left me devastated and completely at a loss as what to do. I have a partial denture at the front but it's far from ideal as it moves around my mouth, even with fix a dent can only be in for a few hours and I can't really eat or drink hot drinks with it. There seems to be a lack of information and I honestly feel like I've been left just to put up with losing all my teeth at 34."*

Assessing a patient's needs at diagnosis, and delivering care in a more integrated way, can help people with brittle bone disease access the specialised dentistry services they need.

Case study 2: Primary immunodeficiency disorders

Care requirements: immunology, rheumatology, haematology, endocrinology, gastroenterology, dermatology, paediatrics, infectious diseases, physiotherapy, mental health support, social care

People with primary immunodeficiency disorders have an immune system that does not work as expected. Not only will people with these disorders suffer from prolonged and recurrent infections such as chest, ear, gastro-intestinal or skin infections; they can also present with blood disorders (e.g. hypogammaglobulinaemia), irregular growth and development, organ inflammation and / or autoimmune disorders (e.g. lupus, rheumatoid arthritis, diabetes). Due to the great variety of symptoms, primary immunodeficiencies can take years to diagnose and require many services to manage. One patient from the Primary Immune-deficiency Patient Support Charity said:

"It took 30 years to receive my diagnosis, and when I did I burst into tears through sheer relief – I knew something wasn't right and that most people couldn't also be feeling the way I was feeling!

"GPs are in some ways the least qualified to diagnose a rare disease, but they have most oversight over the patient's medical history and their condition. We need to find a way of educating GPs or moving complicated diagnoses elsewhere."

For people with primary immunodeficiency disorders, early diagnosis and intervention can help prevent infections that could lead to long-term problems or delay disease progression. Specialised diagnostic services are therefore needed for patients with rare and hard-to-diagnose conditions, such as primary immunodeficiency disorders, and these can be made

more widely available by integrating them into the rapid diagnostic centres / community diagnostic hubs now being established.

The variety of symptoms seen in primary immunodeficiencies also means there is no one-size-fits-all approach to care. Some patients will experience severe physical restrictions and require physiotherapy, occupational therapy and social care support. A holistic needs assessment upon diagnosis as part of an integrated care approach would be beneficial to patients with primary immunodeficiency disorders.

Case study 3: Fibrous dysplasia / McCune Albright Syndrome

Care requirements: paediatrics, orthopaedics, rheumatology, endocrinology, pain management, craniofacial/maxillofacial, ENT, ophthalmology, neurology, physiotherapy, occupational therapy, psychology

Fibrous dysplasia is an incurable, progressive and rare disease that can affect any number of bones in the skeleton, including the spine and the skull. Each patient is unique and the severity of the disease varies: symptoms can go unnoticed for years, or they can start very early on in life and result in significant physical impairment, bone deformity, frequent fractures and chronic pain. Fibrous dysplasia can present as part of a syndrome called McCune Albright Syndrome, which can have endocrinological symptoms including precocious puberty, hyperthyroidism, low blood phosphorus, excess growth hormone, and others. Fibrous dysplasia is therefore very difficult to treat and the Fibrous Dysplasia Support Society UK reported to us that patients have complex needs that are not always met by the current system. For example:

- Due to the large number of health specialisms required, patients commonly have to travel to different hospitals to receive treatment, with communication and record-sharing between them being poor – and with patients feeling distressed, confused and unsure that they are being provided with the correct treatment as a result
- Some patients are discharged and left in the care of their GP and can go without seeing a specialist for many years. However, if a bone is broken or fractured, it needs to be repaired by a specialist who has experience in treating rare bone diseases and many local hospitals do not have this. In addition, patients will require regular specialist blood tests for hormone glands, and these should be checked by a specialist
- The transition from child to adult services can be difficult for patients. Many are not transferred from a children's hospital to adult specialists and they end up without the care they need
- As well as physical health, patients require mental health support to help cope with the chronic pain they experience; with bullying commonly reported by patients with noticeable changes to physical appearance; and with isolation, anxiety and depression.
- Patients also report not having access to wellbeing and community services such as carers, wheelchairs, or alternative therapies

Joining up a wide range of health and care services, including child and adult services – with shared access to patients' medical histories and test results – can have a significant, positive impact on patients with fibrous dysplasia.

Case study 4: Chronic kidney disease

Care requirements: renal services, surgery, immunology, endocrinology, mental health support, dietary advice, physiotherapy, social care

Unlike the other conditions described here, chronic kidney disease (CKD) is common, affecting around 3 million people in the UK. However, CKD is also complex and requires a range of services at different stages of the diseases, and some services – notably renal dialysis and kidney transplant – are specialised NHS services. A CKD diagnosis can have huge implications for a person's quality of life. Patients commonly report sleep disturbances, food and drink restrictions, unemployment, sexual problems, changes in body appearance, limitations in leisure activities and vacations, increased dependence; and uncertainties about the future.[i,ii] Progression to kidney failure can mean facing lengthy hospital visits three times a week to receive dialysis and potentially being placed on the transplant waiting list with the possibility (but no guarantee) of receiving life-saving surgery. Nearly half of dialysis patients experience some form of distress.[iii] In transplant patients, depressive symptoms have been shown to increase the risk of death by 65%.[iv] Yet, currently, mental health support is not a standard part of kidney patient care. SHCA member Kidney Care UK has been campaigning on this issue for some time.

Kidney patients should have mental health support integrated into their routine care to help manage the psychological impacts of their diagnosis, with clear benefits to their physical health.

Unlike the other conditions detailed in this paper, CKD is also preventable. People with high blood pressure, high cholesterol or diabetes are at risk of developing CKD. These risk factors are more common in people from Black or South Asian communities, who are five times more likely to develop CKD and three to five times more likely to progress to kidney failure than other groups. Controlling blood pressure can delay or even prevent the onset of CKD, but only if the person knows and understands their risk and has access to the necessary support to do something about it.

Closer interactions between the health service and local governments – with their experience in public health planning for the local community – could lead to CKD prevention initiatives tailored to at-risk communities.

Case study 5: Stem cell transplant patients

Care requirements: haematology, oncology, surgery, immunology, mental health support, dietary advice, physiotherapy, social care

People who receive a stem cell transplant to treat blood disorders or cancer have significant and complex needs relating both to their disease and the effects of the treatment itself. The side effects of a stem cell transplant can last for several months and even years. Access to holistic care and support is crucial to improve survival and quality of life. Some examples include access to nutritional advice and physiotherapy to manage the side effects of graft versus host disease, access to secondary cancer screening, or help to cope with physical, emotional and financial changes.

Unfortunately, the way that these services are currently commissioned and reimbursed often leads to disjointed care and variation across the country. Services are either the responsibility of NHS England or CCGs, and are delivered in a variety of settings – with the lack of integration often adversely impacting care. The charity Anthony Nolan has found that patients have to repeat the same information to multiple services, feel as though they are cut off from any support after their discharge from hospital, and miss out on vital services. One patient said:

"*When I first left hospital I felt so alone, more or less dropped from a great height and left to deal with it. This isn't how it should be. I feel passionately that the whole person needs to be cared for and not just their illness*."

People who have had stem cell transplants require care that is planned around their needs. An integrated system could and should deliver seamless transitions between care settings, shared data, holistic support encompassing their clinical, emotional and financial needs, and an emphasis on early intervention.

Case study 6: Huntington's disease

Care requirements: Social care, Neurology, Psychiatry, Psychology, Physiotherapy, Speech and Language Therapy, Dietetics, Occupational Therapy

Huntington's disease is a complex, hereditary, neuro-psychiatric disease that affects all aspects of an individual's life and is incredibly challenging for the family who are supporting them. Huntington's is also a progressive illness. People don't fit neatly into 'physical health needs', 'mental health needs' or 'social needs' therefore teams need to work together to look after the individual. People with Huntington's require continuity of care and a named professional who can co-ordinate that care – and this is not always available in the current system, and both patients and carers suffer as a result.

One carer said: *"Initially my husband had a named social worker. She was with us for 10 years, she knew my husband well and he trusted her, we knew who to go to as issues arose. Things changed when she retired and since then we have been passed from pillar to post. He has been seen by a variety of teams – the complex care team, community health team, social services, mental health team and the dementia service. He is on a caseload and then the case is closed after 6 weeks, we then have to go through the GP for a new referral. Each team seems to pass to the next rather than having any continuity. In a crisis we are told to go to A&E. Our latest crisis was when he tried to take his own life (he has done this four times). The police attended but there were no beds available and he stayed in the community. I battled for the mental health team to assess him. I was told he needed a placement but, again, there were no beds. I did the research and found him a suitable care home. I had to fight to get him there and to keep him there (I was told he would be moved after 2 weeks because it wasn't council run). What I need, as the wife and carer of someone with Huntington's disease, is continuity, for teams to work together. My husband wants to come home and I want him home. But to do this we need a care package and a team to go to if a crisis arises. I am exhausted and my own health has suffered significantly. Every step is a battle."*

The Neurological Alliance works in partnership with more than 80 organisations, including the Huntington's Disease Association, to campaign for greater awareness and understanding of neurological conditions and to ensure that every person diagnosed with a neurological condition has access to high quality, joined up care and information from their first symptoms, throughout their life.

Recommendations

Given the above, we make the following recommendations to ensure moves towards integration deliver genuine improvements in standards of care.

In order to address inequality and unwarranted variation, and improve care standards:

- **National standards of care** must be implemented (where they do not already exist) and adhered to, which is particularly critical if integrated care systems (ICSs) are to take on more responsibility for specialised commissioning. These could include minimum

standards of care, including access to clinical nurse specialists, as well as requirements to complete holistic needs assessments and develop comprehensive care plans

- **Budgets should be delegated only with adequate safeguards**. To help join services together, a single budget holder in an integrated care organisation may be beneficial, but only if they have the expertise and capacity to deliver specialised services, including where patients are likely to travel outside of ICS borders to receive care
- **The patient voice must be embedded in integrated care organisations**. To ensure the voice of people with rare and complex conditions is not lost as decisions are taken on changing pathways, patient and public involvement is critical
- **Innovative and effective treatments should be provided quickly and on a uniform basis across the country**. The needs that people have, rather than the place they live, should determine what clinicians are able to offer by way of treatment
- **Healthcare professionals should be increasingly aware of the needs of people with rare and complex conditions**. If integrated services are to become more widespread, it is important they address the needs of people with both common and less common conditions
- **Rapid Diagnostic Centres / Community Diagnostic Hubs** should be expanded to ensure patients with unexplained symptoms that are not necessarily linked to cancer receive faster diagnoses wherever possible. Often GPs will 'guess' based on the symptoms and send people to many different specialists
- **People who do not have a confirmed diagnosis should be directed to the local genetics service** in areas where diagnostic hubs are not yet established to determine whether it is possible for them to gain a diagnosis. Local genetics service can act as a hub to diagnose people and have the knowledge needed to direct them to the local specialist clinic that is most likely to be able to support them effectively based on their symptoms
- **Minimum waiting time standards** should be created for people with rare and complex conditions to support more rapid diagnosis and better access to specialist care

In order to support holistic care planning:

- A **holistic needs assessment should be performed at the point of diagnosis** for patients with rare and complex health conditions
- A **comprehensive care plan should be decided jointly between the patient and their medical teams** following a needs assessment, which should include a **detailed explanation of the services they will be referred to and equipment they need** to support their overall health, mental health and quality of life. All services should be included in this plan, including dentistry, physical and psychological therapies
- People with rare and complex conditions should be provided with access to a **named clinical nurse specialist** to provide greater continuity and coordination of care
- A patient's care plan should be **reviewed at regular intervals throughout the patient's life** to ensure their needs are being met, and if **new technology** exists that could better support the patient's needs
- Consideration should be given to the **cost effectiveness of investment in technology**, such as a specialised electric wheelchair, which improves a patient's quality of life and therefore overall health, as well as reducing social care needs, when making decisions about care
- **Children with rare and complex conditions must be prepared for the transition from child to adult services in advance**, and the transition should be carefully managed so that all the required adult services are made available to the patient. Children should be allowed to meet their new adult medical team in advance of the transition and a new holistic needs assessment should be performed

- **All patients, young and old, with chronic conditions should be offered mental health support**

In order to support better communication and information regarding rare and specialised health conditions:

- Different providers must have access to the information they need about a patient or they will not be able to make informed decisions about the next steps for a patient's treatment or care, and this may require **improvements in IT infrastructure**. There is often **distress and confusion in attending different hospitals** for appointments as there is **poor communication** between hospitals and other parts of the system. Our members report that patients themselves often have to ask for copies of scans so they can take them along to different appointments
- An **online information resource on rare diseases**, accessible to all providers of health and care services – especially in primary care – should be made available to improve recognition of symptoms, or disease progression, to aid faster diagnoses and treatments decisions. This information should include prevalence and severity of the condition, and whether specialist services to support patients with the condition should be commissioned by NHS England centrally or at ICS level

Conclusion

This short briefing note is intended to illustrate – through the use of case studies – the importance of integrated services for patients with rare and less common conditions. It also sets out a number of recommendations which may be beneficial for those planning the integration of services, particularly for those involved in establishing the new 'integrated care systems' in England.

For any further information, please get in touch with team@shca.info.

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