Setting goals of care in acutely unwell patients with chronic neurodisability

In the broadest terms, the 'goals of care' are the common purposes towards which clinicians, patients and their family and carers are working. Examples of goals of care include curing a curable disease (e.g. pneumonia), reducing seizure frequency in an individual with epilepsy, or reducing painful symptoms in an individual with inflammatory arthritis.

The phrase 'goals of care discussion' has often been used in a narrower sense, focusing on end-of-life care planning and decisions around the degree of resuscitation appropriate for an individual in the event of a cardiorespiratory arrest. This is a reductive view of goals of care discussions, which can, and should, serve a much broader purpose.

Open discussions around goals of care should enable patients and their family and carers to share their values, hopes and fears, and clinicians to provide information regarding the diagnosis, prognosis and guidance on available resources (Vaughan and Kluger, 2018). Goals of care should be considered dynamic and open for regular review. Consequently, goals of care discussions should be seen as an ongoing dialogue with patients and their family and carers, and not as entirely isolated meetings with binary outcomes with regards to decisions around resuscitation. Appropriately, the priorities of patients and their family or carers will change over time, as health conditions progress and life circumstances change. This is particularly the case for children and young people approaching adulthood, passing through the process of transition between paediatric and adult services.

Patients with neurodisabilities

The term 'neurodisability' describes a group of congenital or acquired long-term conditions that are attributed to impairment of the brain and/or neuromuscular system and create functional limitations (Morris et al, 2013). Even when the result of a static insult to the brain (e.g. traumatic brain injury), functional limitations may vary over time, and can occur in isolation (e.g. motor difficulties following brachial plexus injury) or in combination. Difficulties may include abnormalities of movement and tone, cognition, hearing and vision, communication, emotion and behaviour (Morris et al, 2013).

Cerebral palsy is the mostly common cause of neurodisability in the young encountered in clinical practice, and so is often used as an exemplar condition. The term encompasses a range of disorders which result in a non-progressive injury to the developing brain (<2 years of age), causing abnormalities in motor function or development, and may be associated with other problems (Colver et al, 2014). Cerebral palsy affects 2–3.5 live births per 1000 in the developed world.

Children and young people with cerebral palsy have more hospital admissions, longer hospital stays, more investigations and more diagnosis then other children and young people (Murphy et al, 2006). The 2013 Royal College of Paediatric and Child Health report 'Overview of child deaths in the four UK countries' demonstrated that 71% of children who died had chronic conditions, most frequently neurological, pointing to the need to focus on quality of care in children with chronic conditions, particularly neurodisability. This also highlights the unavoidable fact that some children and young people with neurodisability will die before reaching adulthood.

Life expectancy in individuals with cerebral palsy can be related to the severity of mental, manual, ambulatory and visual impairments, with only marginal reduction if only mild impairment is seen in all of these domains (Colver et al, 2014). In a large cohort of children in the UK with cerebral palsy alive at 2 years of age, who had severe impairments in all four of these domains (mental, manual, ambulatory and visual), 72% lived to 10 years of age, 44% to 20 years of age, 34% to 30 years of age and 27% to 40 years of age (Colver et al, 2014). Life expectancy with other causes of neurodisability may be much less predictable, particularly when the underlying disorder is progressive.

Planning and care for children at risk of premature death needs to improve (Horridge, 2015). The National Confidential Enquiry into Patient Outcome and Death (2018) chronic disability study was undertaken with the aim of identifying the remediable factors in the quality of care provided to children and young people with chronic disabling conditions, focusing in particular on cerebral palsy. The findings were presented in the report *Each and Every Need*, with 35 recommendations to improve the quality of care for children and young people with neurodisability (the nine recommendations pertinent to acute admissions of patients with neurodisability are presented in *Table 1*).

Even if not working in the field of neurology or neurodisability directly, junior clinicians working in an acute inpatient setting will almost inevitably encounter patients with neurodisability. It can be daunting to become responsible for the day-to-day care of a patient (child or adult) with a chronic neurodisability for the first time. Communication can at first be challenging, with uncertainty as to levels of understanding and/or cognition. There may be a constellation of involuntary movements and postures, and a wide range of comorbidities requiring management (bringing with them a potential cornucopia of familiar and less familiar medications, along with multiple other teams with whom care will need to be coordinated). It is important to ensure at all times, even if inadvertently, that a patient's neurodisability per se is not used as a reason to deny him/her treatment which is potentially available to any other patient, in line with the Equality Act 2010 in the UK.

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Table 1. Recommendations of the *Each and Every Need* report relating to acute admissions of patients with neurodisability

All patients with complex needs and, where appropriate, their parents, carers or legal guardians, should be offered the opportunity to develop a patient-held emergency health care plan or emergency care summary to facilitate communication in the event of a health-care emergency	
This should include as a minimum:	1. Information about the patient's health conditions and treatment
	2. Who to contact in a range of scenarios and what to do
	3. A statement about what has been discussed and agreed about levels of intervention including palliative care planning
	4. The existence of any advance directives (for those over 18 years), lasting power of attorney or any other measure
The existence of this emergency health care plan or emergency care summary must be recorded in all communication and case notes and this should be subjected to local audit	
Patients with a neurodisabling condition should have an assessment completed by their lead clinician to determine their risk of respiratory compromise. This should be reviewed as appropriate for the complexity of the patient's needs. Those patients at significant risk of respiratory compromise should be assessed by clinicians with expertise in respiratory medicine, in order to discuss with the patient and family the range of interventions most likely to lead to the best outcome. 'What to do' and 'who to contact' in the event of respiratory symptoms should be documented in the patient-held emergency health care plan	
As for all patients, those with a neurodisabling condition admitted to an acute general hospital as an emergency should have timely assessment and senior review within 14 hours of admission by a specialist relevant to the emergency as recommended by the Royal College of Paediatrics and Child Health (2015a,b) and the Royal College of Physicians (2012)	
Pain scoring tools should be understood and used in the perioperative and periprocedure period for patients with a neurodisabling condition. Health-care staff should be trained in their use	
All medically frail patients with a neurodisabling condition and, where appropriate, their parents, carers or legal guardians, must be offered the opportunity to discuss their care wishes in the event of serious illness or sudden collapse with their lead clinician. This should be recorded in their patient-held emergency health care plan. This may include discussing do not attempt cardiopulmonary resuscitation decisions and palliative care plans, which should be validated at each point of care according to the existing legal requirements and professional guidance. This is particularly important to have in place at handover during transition to adult services	
Each consultation with patients with a neurodisabling condition should be used as an opportunity to enquire whether they and their family have the information and support they need	
Clinicians should be aware of, and comply with, the ethical and legal requirements for consent to surgery as defined by the General Medical Council (2008) and requirements for mental capacity assessments which will vary depending on which UK country they live in. These requirements must be communicated clearly to patients, parents and carers and documented in the case notes	
Patients with a neurodisabling condition should be involved in all communications and decision-making about their care and management where possible, and where appropriate, with adjustments in place to support their involvement, including specialist speech and language therapists as required. Parents, carers or legal guardians must also be included in these conversations as appropriate	
After a period of inpatient care patients with a neurodisabling condition should have their ongoing function and daily needs assessed and documented. Any significant change which would necessitate a planned alteration to day-to-day care must be clearly communicated in discharge plans. The discharge plan should be sent to the patient, the parents or carers, and the multidisciplinary team, including the GP	

From National Confidential Enquiry into Patient Outcome and Death (2018)

Considering the needs of patients with neurodisability during an acute admission

The first contact that an individual clinician has with a given patient with neurodisability may be during an acute admission with a medical problem. This may be the development of a new health issue (e.g. acute appendicitis), or an acute exacerbation of a chronic issue relating to the underlying neurodisability. It is important to establish the goals of care for that given admission, within the overall context of the goals of care for that individual.

The fundamental needs of patients with neurodisability are no different than for any other patient on the ward and are well conceptualized through Maslow's hierarchy of needs (*Figure 1*). The most basic level of need is the physiological – e.g. breathing, food, sleep – and while ensuring provision for needs may be more of a challenge for the patient with neurodisability, it is essential that elements higher up on the hierarchy are not forgotten.

Traditionally, the focus of clinical encounters is around the diagnosis and treatment of chronic disease and disorders, with less focus on functional abilities and participation in society. However, the World Health Organization (2001) International Classification of Functioning, Disability and Health (*Figure 2*) can be used as a framework to conceptualize disability and health. While often used in the context of an outpatient appointment, this is an excellent frame of reference when approaching any clinical encounter with an individual with a neurodisability, although acute inpatient encounters are necessarily likely to focus upon body functions and structure (the level of impairment).

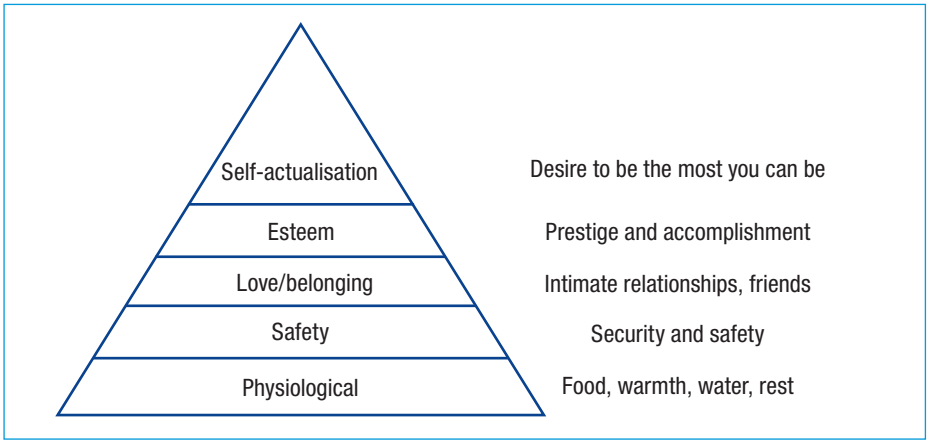

Figure 1. Maslow's hierarchy of needs.

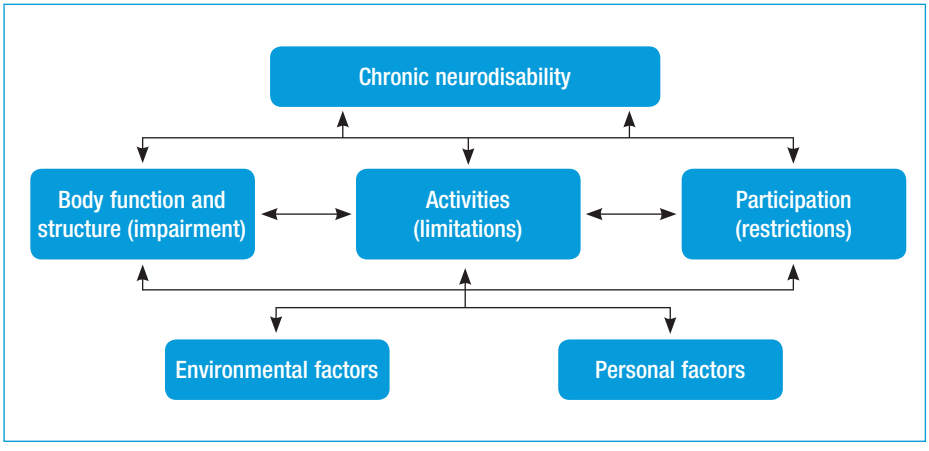

Figure 2. The international classification of functioning, disability and health. From World Health Organization (2001).

Get to know the patient and the care team surrounding them

Complex patients typically require complex care, often delivered by a broad team of clinicians, allied health professionals and social care professionals. When meeting a patient with neurodisability for the first time it is important to take the time to establish key facts about the patient, and the teams involved in his/her care. It may be the case that the individuals accompanying the patient are not the best placed to provide a clear background beyond the immediate acute presentation. It is essential to establish early the patient's level of cognition and understanding (which of course may be entirely normal even in the face of very profound communication difficulties), details of where the patient resides, and who is involved in his/her daily care. A full list of professionals involved in his/her care should be established, particularly as they may need to be contacted directly as an admission progresses. Many patients with neurodisability will have a key worker who will be an ideal point of contact to provide these details. For children and young people admitted to hospital, it is also important to establish who has parental responsibility (see below).

Easy to overlook in the middle of a busy acute shift, but just as important is to get to know the patient as a person – what are his/her likes and interests? An individual with a neurodisability is not defined solely by his/her neurodisability. A good starting point with this is to establish what name the patient would like to be addressed by. Be sure not to forget to include this information in your handover of the patient during shift changes.

Issues of consent for patients with neurodisability

It is the clinician's duty to ensure that they have a patient's consent, or other valid authority, before treating them. The General Medical Council (2008) provides useful guidance around issues of consent in patients with potentially diminished capacity. The key points of this may be summarized as:

- A physical examination must be carried out if indicated – do not avoid it because of difficulties in communication or assessing capacity. Instead try to gain the patient's trust and persuade him/her to consent to an examination
- Capacity is decision-specific and time-specific
- Every attempt must be made to maximize a patient's capacity to make a decision, including providing information in a way the patient can understand
- Doctors must work on the presumption that every adult patient has the capacity to make decisions about his/her care
- Lack of cooperation should not be interpreted as valid refusal of consent, without first assessing capacity
- Doctors should explain, in a way that the patient can understand, the implications of a refusal to cooperate.

If an adult patient with neurodisability lacks the capacity to make decisions about his/her care, then it is possible that he/she will have authorized someone over 18 years of age to make decisions for him/her under a lasting power of attorney, or that the Court will have appointed a deputy to give consent. If no such arrangement is in place, then decisions must be made on the basis of the best interest of the patient. The General Medical Council (2008) again provides helpful guidance for this:

1. Make the care of the patient the first concern
2. Treat patients as individuals and respect their dignity
3. Support and encourage patients to be involved, as far as they want to and are able, in decisions about their treatment and care
4. Treat patients with respect and do not discriminate against them.

Children under 16 years of age can consent to their own treatment if they are considered to have enough intelligence, competence and understanding to fully appreciate what is involved in their treatment. If these conditions (frequently termed 'Gillick competence') are not met, then consent can be provided by an adult with parental responsibility (which may include either parent, a legally appointed guardian, a person with a residence order concerning the child,

a local authority designated to care for the child, or a local authority or person with an emergency protection order for the child). Urgent, life-saving care may be delivered without explicit consent.

Practical issues of care and symptom management

Some practicalities of day-to-day care of patients with neurodisability, particularly around symptom management, are outlined in *Table 2*.

Considering limitations of care

During an acute admission with a significant illness for a patient with neurodisability, inevitably the issue will arise as to what should be done if a significant deterioration in medical condition occurs. It is all too easy to approach this in a fairly binary fashion (i.e. should cardiopulmonary resuscitation be performed in the event of a cardiorespiratory arrest? Should this individual be admitted to the critical care ward in the event of a deterioration?). Discussion around these end of life issues is much more nuanced, and an emphasis should be maintained on the active management of symptoms in line with the overall goals of care for the patient. For some patients an advanced care plan may already have been prepared, but for others there may have been no such discussions.

A review and synthesis of best practice for goals of care discussion identified that common pitfalls or failings are that they are often conducted by physicians who do not know the patient, do not routinely address patients' non-medical goals, and often fail to provide patients with sufficient information about prognosis to allow appropriate decisions (Bernacki et al, 2014). Best practice included the sharing of prognostic information, eliciting decision-making preferences, understanding fears and goals, exploring views on trade-offs and impaired function, and wishes for family involvement (Bernacki et al, 2014).

A key consideration is who should attended the goals of care discussion, not just in terms of members of clinical staff, but also which family members and carers. Where ever possible, the goals of care discussion should be postponed until all key individuals are able to attend. Wherever possible, the discussion should be led or chaired by a clinician who has been involved in the long-term care of the patient.

Table 2. Practical issues to consider during acute admissions of a patient with neurodisability

Issue	Consideration
Abnormalities of tone or posture	Dystonic posturing may be mistaken for generalized seizures, leading to excessive and unnecessary treatment. Tone difficulties are likely to be exacerbated during acute illness and may require expert management from an early stage. This may further impact upon communication and feeding
Bladder	Urinary retention may be precipitated by or may precipitate an acute admission. Managing clinicians must be particularly vigilant for retention in patients for whom communicating this problem may be challenging. This may worsen pre-existing problems with abnormally elevated tone
Bowels	Constipation is a common problem in acutely admitted patients, more so in the context of a chronic neurodisability when many anti-tone medications may adversely affect gut motility. Constipation may drive significant pain and exacerbate dystonia, and additionally reduce appetite and feed tolerance
Communication	This is a common difficulty in patients with chronic neurodisability, which may be compounded by visual and/or hearing problems. Do not presume this means problems with cognition. When difficulties with verbal communication exist, augmentative and alternative communication includes any 'add-on' to speech, including gestures or signing, facial expression, eye gaze, switching and more advanced augmentative technologies. Acute illness may further restrict access to augmentative and alternative communication, adding further difficulty for an individual during an acute admission
Discharge planning	This should begin at the point of admission, as it may potentially be highly complex, particularly if the care needs of a patient are likely to have substantially changed at the time of discharge. Lack of forward planning for discharge is likely to unnecessarily prolong admission. An early multidisciplinary team involving clinicians involved in ongoing care as well as those managing the acute episode is to be encouraged
Early warning systems	These are typically validated in population samples of otherwise healthy children and/or adults. Their validity is uncertain in patient groups whom may have abnormalities in baseline physiological parameters
Feeding	This should not be considered a binary choice – i.e. the patient is or is not safe to feed. A much more nuanced situation is likely to be encountered, e.g. thickened but not un-thickened fluids are safe orally. Feeding difficulties may worsen in acute illness, and so early review by a speech and language therapist should be encouraged
Medications	Polypharmacy is commonly experienced by patients with chronic neurodisability. Care must be taken when adding additional medications during acute admissions for their potential to interact with baseline medications
Pain	This is a commonly experienced problem in people with motor disabilities. Ability to communicate the extent of pain may be further limited during an acute admission, and so managing clinicians must be vigilant and proactive in assessing pain
Seizures	These are a common problem in patients with neurodisability. Many individuals will have a bespoke seizure plan differing from those in Advanced Paediatric Life Support or Advance Life Support guidelines. Plans should be in place for managing clustering of seizures, as well as single prolonged seizure episodes

The output of the goals of care discussion is likely to differ between clinical settings and institutions. A written care plan is likely to be produced, which may take the form of:

- A symptom management plan
- An advanced care plan
- A statement of wishes and preferences.

Guidance for preparation of these documents (and the discussions leading up to this point) for children and young people has been developed by Working Together for Short

KEY POINTS

- Patients with neurodisability, particularly children and young people, experience more acute hospital admissions then their otherwise healthy peers.
- Cerebral palsy is the most commonly encountered cause of neurodisability in the young, and has been used as an exemplar condition to explore the quality of care received by patients with neurodisability.
- Recommendations for improving the care received by patients with neurodisability have recently been published by the National Confidential Enquiry into Patient Outcome and Death report *Each and Every Need*.
- The goals of care are the agreed aims towards which professionals, patients and their family and carers are working.
- Goals of care discussions between professionals, patients and his/her family or carers are not solely focused upon decisions around end of life care, although these are important considerations.
- Goals of care discussions should be considered a dynamic process, revisited as the priorities and needs of the patient and his/her family or carers changes over time.

lives (http://www.togetherforshortlives.org.uk) and the Council for Disabled Children (http://councilfordisabledchildren.org.uk), and for adults by the National Council for Palliative Care (http://ncpc.org.uk). The General Medical Council (2010) also provides an ethical framework with specific guidance on treatment and care towards the end of life which all UK clinicians are bound to follow.

In many clinical institutions, if a decision has been reached to limit the degree of resuscitation to be attempted in the case of a cardiorespiratory arrest, then documentation of a formal 'do not resuscitate' order will be required in the patient's medical notes.

The importance of reflection

Caring for a patient with a chronic neurodisability is very often extremely rewarding, but with increasing clinical complexity comes increasing opportunity for shortfalls in care to arise. Even when a high standard of care has been delivered, there is often limited capacity for modern medicine to modify an underlying disease process and ameliorate the burden of disability, or to ensure a continuing good quality of life as primary or secondary consequences of a disorder progress. More so perhaps than with other patients, it can be an emotional experience caring for patients with neurodisability. It is important to take the time to acknowledge this, and to reflect on the experience of providing care during an acute admission, both in regards with what went well and what could have been done better. **BJHM**

Conflict of interest: none.

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