

Care Pathway Mapping for Dravet Syndrome (DS) Patients in England – interim results from a pilot study

Purpose

Dravet syndrome (DS) is an epileptic encephalopathy estimated to affect 1 per 15,500 live births [1,2], characterised by seizures that start in the first years of life, triggered by fever, that persist and are usually multi-drug refractory. Finding effective treatments is imperative to reducing seizures and improving patient outcomes: DS is highly burdensome for both the patient and their family as well as the healthcare system.

The National Institute for Health and Care Excellence (NICE) guidelines [3] provide a recommended care pathway for general epilepsy in paediatric and adult patients. However, there is sparse detail on the specific management of patients with rare genetic epilepsies. Recommendations include:

- "When a child presents with suspected DS this should be discussed with, or referred to, a tertiary paediatric epilepsy specialist."
- "When presentation includes behavioural or developmental regression, this should result in immediate referral to tertiary services."
- "All epilepsy patients should have one or more reviews by a specialist annually."

It is important to understand how care is currently delivered to children and adults to improve care, so that it is comprehensive and meets the needs of DS patients. We therefore aimed to:

- Describe how care for people with DS is delivered across England
- Determine the utilisation of healthcare resources for the treatment of patients with DS, and the effects of seizure burden and age group.

Methods

1: Systematic Literature Review (SLR)

- A comprehensive SLR was conducted to find and assess published studies that reported on healthcare resource use and/or costs for patients with DS, and specifically to identify UK data.

2: Interviews with clinicians

- The SLR identified evidence gaps for emergency and ongoing management resource use, and additional information was required to understand the patient pathway and how care is delivered for DS patients. Therefore, we conducted interviews with epilepsy clinicians.
- We developed a discussion guide and conducted semi-structured pilot interviews with three experts to map out DS patient care and map an initial version of the care pathway.
- A purposive sampling approach was taken to recruit an additional 15 clinicians directly involved with the care of children and adults with DS in England to further understand how care is delivered and to finalise the care pathway for individuals with DS (Figure 1).

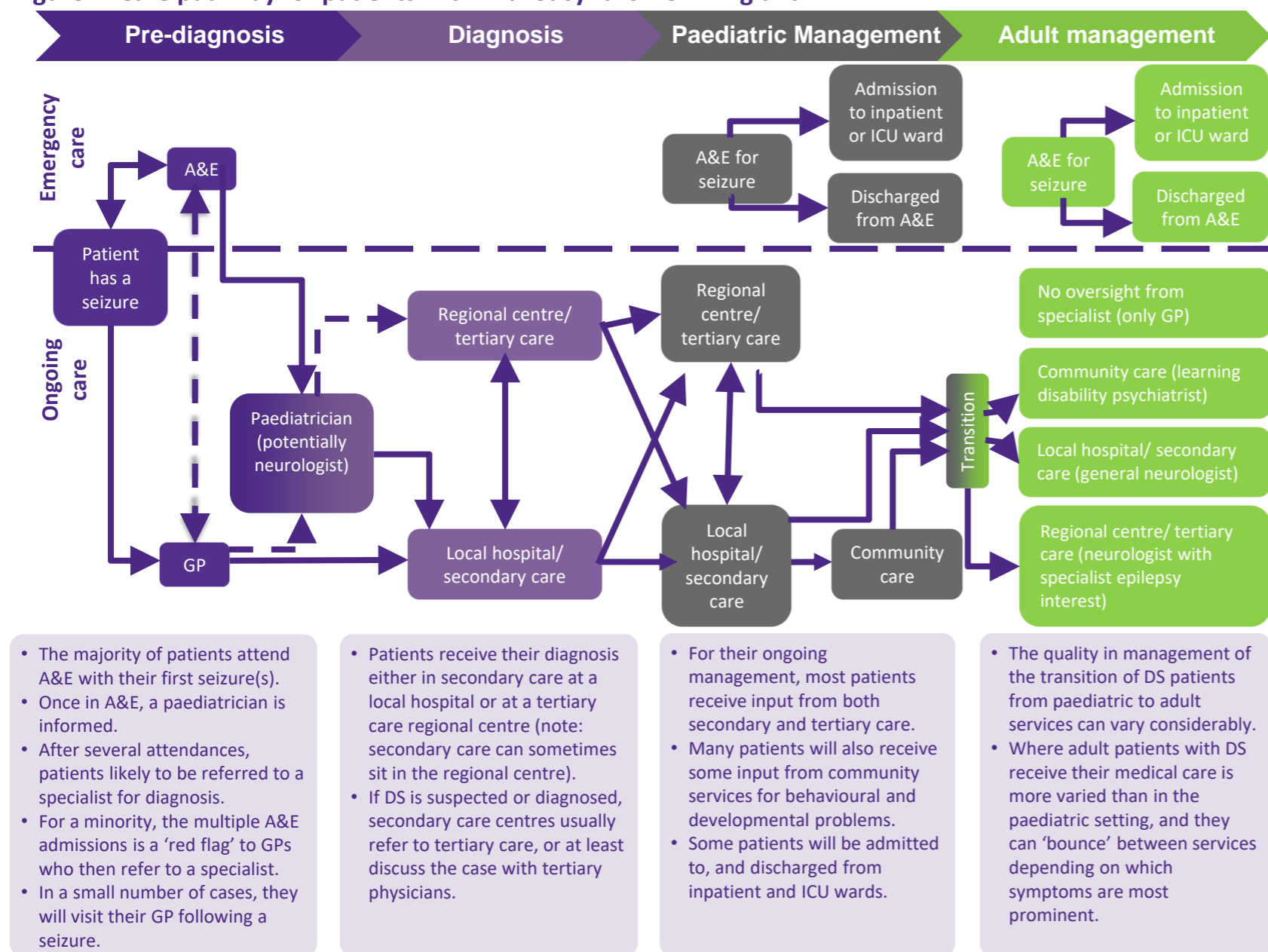
3: Validation exercise

- To validate the research findings across the collective sample, additional specific information and clarification on resource use were required. A formal validation exercise was developed by creating a questionnaire with data tables for the previously interviewed participants to complete. Nine out of 16 participants from Phase 2 sent back the validation exercise for Phase 3.

Results

Dravet Syndrome Care Pathways and Variations in Archetypes of Care

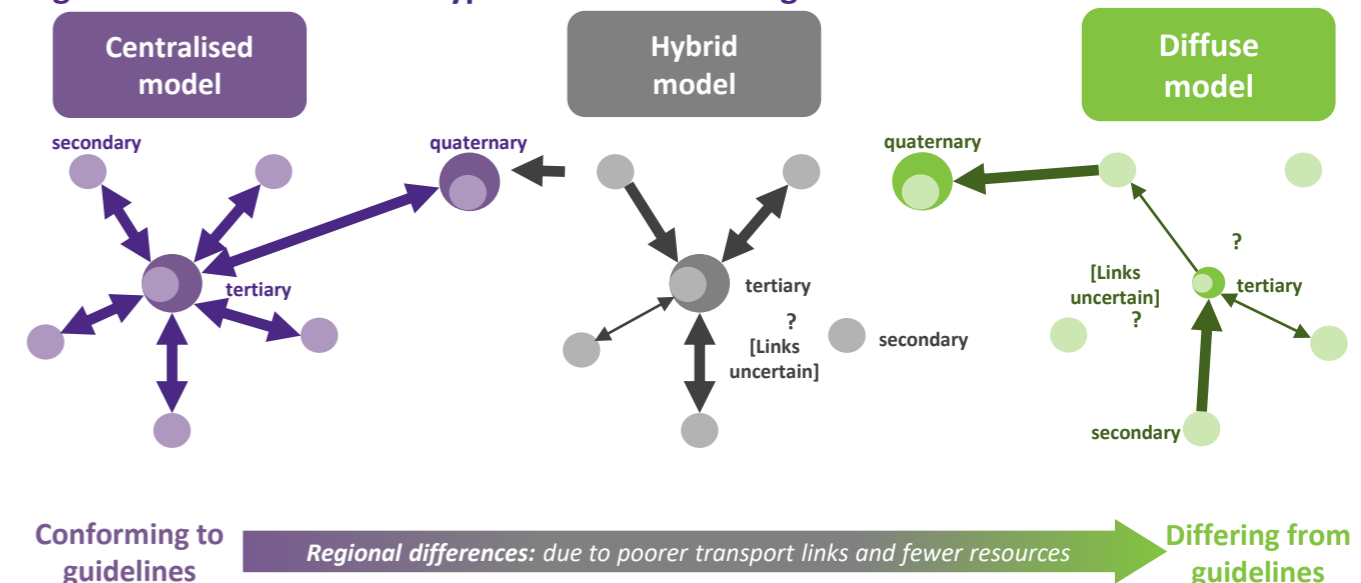
Figure 1. Care pathway for patients with Dravet syndrome in England



In comparison to the published NICE pathways for general epilepsy in paediatrics and adults [4], the DS care pathway (Figure 1) varies in practice and in detail, particularly in terms of consistency of care compared to the NICE recommendations. Paediatric services were more likely to follow a 'joined-up' approach, in contrast with adult services, while in many cases transitioning arrangements between paediatric and adult services were lacking.

Data also showed regional variation in care and service provision of paediatric services across England. Three main archetypes of care were described by the interviewees: 'centralised', 'hybrid' and 'diffuse' models (Figure 2). The 'centralised' model conforms closer to the NICE guidelines, while the 'diffuse' model is more likely to differ from the recommendations, particularly that there are poorer links between secondary and tertiary services. The non-homogeneity was attributed to regional differences in transport links and availability of healthcare resources.

Figure 2. Variation in archetypes of care across England



Resource Use and Costs

SLR and clinician interviews show the high burden of resource use and costs which varied across age, seizure frequency and services. Table 1 shows clinician reports on the variation in emergency resource use after receiving rescue medication by patient age group. Younger patients have a higher demand on ambulance and A&E services, while older patients who attend A&E are more frequently admitted to hospital. Table 2 shows the variation in the frequency of ongoing resource use by seizure frequency by type of provider (note: quaternary care is specialised tertiary care specific to DS). Average convulsive seizure frequency per month was defined by clinicians for paediatric and adult patients in the validation exercise. High seizure frequency patients had much higher ongoing resource use compared to medium and low seizure frequency patients, particularly for secondary care and nurse telephone and email consultations.

Table 1. Annual emergency resource use after rescue medication use, by age group (average)

Patient age group (years)	Percentage of those who call an ambulance after administration of rescue meds	Percentage of those who called an ambulance that attend A&E	Percentage admitted from A&E to the:	
			Hospital	ICU
2-3	82.3	76.3	55.0	18.0
3-5	63.8	56.3	45.0	14.3
5-8	47.5	42.5	43.8	14.3
8-14	27.5	37.5	36.3	11.8
14-18	26.7	43.3	21.7	2.3
18-25	26.5	74.0	71.0	12.0
25+	24.5	75.8	71.0	12.0

Table 2. Annual average frequency of appointments (face to face (F2F) or phone/email consults (non-F2F), for patients with high, medium and low convulsive seizure frequency, by age groups. (NR: Not reported in validation exercise)

Resource Use (avg. appts./yr.)	Convulsive Seizure Frequency (per month)																							
	High								Medium								Low							
	2-3	3-5	5-8	8-14	14-18	18-25	25+	2-3	3-5	5-8	8-14	14-18	18-25	25+	2-3	3-5	5-8	8-14	14-18	18-25	25+			
Community	1.5	1.5	1.5	1.7	2.3	6.0	6.0	1.3	1.3	1.3	2.0	2.0	4.0	4.0	1.5	1.5	1.8	2.2	2.2	1.0	1.0			
Primary	1.3	1.3	1.3	0.7	0.7	6.0	6.0	0.8	0.8	0.8	0.7	0.7	2.0	2.0	0.7	0.7	0.7	0.7	0.7	1.0	1.0			
Secondary	<i>Doctor (F2F)</i> <i>Nurse (F2F)</i> <i>Doctor (non-F2F)</i> <i>Nurse (non-F2F)</i>																							
Tertiary	<i>Doctor (F2F)</i> <i>Nurse (F2F)</i> <i>Doctor (non-F2F)</i> <i>Nurse (non-F2F)</i>																							
Quaternary	<i>Doctor (F2F)</i> <i>Nurse (F2F)</i> <i>Doctor (non-F2F)</i> <i>Nurse (non-F2F)</i>																							

Conclusions

- Patients with DS and their carers have substantial care requirements. The current patient pathway in England is complex, with clinicians reporting substantial variability in the quality of how patients are managed and inconsistencies with the NICE recommendations.
- To prevent inequalities in the care received, there is a need to reduce regional variation in service provisions for DS patients, and to improve transitioning arrangements into adult services. High resource use may equate to high seizure related risks, which can be mitigated with effective treatments.
- This study found a high burden of ongoing and emergency healthcare for DS patients in England, which varies depending on age, seizure severity and access to services. This has not been reported previously to this level of detail, and can help decision-makers understand the high burden of care required by DS patients, their primary care-givers and the broader family unit.
- In many DS patients there is a high unmet need for new therapies that reduce the frequency of convulsive seizures to reduce the burden on and associated cost of healthcare services.

References and Disclosures

References

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