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Title

Benign Childhood Epilepsy with Centrotemporal Spikes (BECTS) and Developmental Co-ordination Disorder

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Abstract

Background: Benign Epilepsy with Centro-Temporal Spikes (BECTS) is a common childhood epilepsy syndrome also known as Rolandic Epilepsy (RE). Neurocognitive phenotypes have been described with greater focus on attention, reading and language domains but there have been far fewer studies focusing on motor functioning. This study included measures of motor, language and cognition in order to investigate the range, degree and pattern of difficulties associated with BECTS in a case series of children, but with a particular emphasis on motor skills.

Method: Twenty-one children aged between 8-16 years with a diagnosis of BECTS were asked to complete standardised assessments for language, cognition, motor functioning and handwriting.

Results: When measuring across language, cognitive and motor domains, 19 (90.48%) of the twenty-one children with a diagnosis of BECTS showed some difficulties on at least one area of functioning using standardised assessment tests. Of particular note nearly half of the (47.62%) children had some difficulties in one or more areas motor functioning.

Discussion: Children with BECTS have a heterogeneous pattern of neurocognitive impairments. The presence of motor difficulties (DCD) should be considered in all children routinely seen in clinical settings with BECTS and included in any screening processes.

Key Words: Child Psychology, General Paediatrics, Neurodevelopment, Paediatric Practice

1. Introduction

Benign Epilepsy of Childhood with Centro-Temporal Spikes (BECTS) also known as Rolandic Epilepsy (RE) is characterised by childhood focal sensorimotor seizures and characterised by hemi-facial motor seizures and interictal centrotemporal (rolandic) spikes on EEG that are activated by drowsiness or sleep. Onset occurs between the ages of 3 and 13 years, and seizures resolve by age 15 to 17 years. It is the most frequent form of epilepsy and the incidence has been cited as 6.2-21 per 1000 children aged 15 years and younger [1].

BECTS was thought to have a benign outcome. However, a series of studies highlighting a range of neurocognitive defects relating to BECTS have brought this into question. Studies have cited a heterogeneous picture with associations noted for example with specific language impairments [2]; Dyslexia and reading difficulties [3]; attention and concentration and Attention Deficit Hyperactivity Disorder (ADHD) [4; 5]; and Autism Spectrum Disorder (ASD) [6].

A review by Vannest et al. (2015) commented that a number of studies had concluded that “the developmental progression of cognitive and behavioral problems, structural and functional changes in the brain, seizures, and the relationship to CTS [Centro-Temporal Spikes] patterns is unclear” (p.89)[1].

Far fewer studies have focused on the motor domain, despite it being mentioned. Smith et al (2012), for example, discuss a neurocognitive phenotype and motor is noted as an important area, yet they do not use any motor measures in the assessment battery [7]. Vannest et al (2015) also in a review paper discusses that many studies are associated with a number of cognitive domains [1]. However, very few have specifically mentioned Developmental Co-ordination Disorder (DCD). One small study by Scabar et al. (2006) describes the association between DCD and BECTS [8]. Rolandic spikes were noted in 70% of children with severe DCD, and conversely 30% of the children with BECTS met the criteria for severe DCD. Overvliet et al. (2011) in their study of 48 children asked parents about their children’s motor difficulties but did not specifically examine them [9]. They found an association between parents reported motor difficulties and reading difficulties in their children, and noted that problems with motor development were reported in 22.9% of the cohort studied. Reilly et al. (2015) explored children with ‘active epilepsy’ (not specifically focusing on BECTS) using a parent report screening tool for DCD (DCD-Q), and found that 29% of the cohort met DSM-IV-TR criteria for DCD [10].

Recognising the specific patterns of neurocognitive impairment when planning support for the individual child, both in the short and longer term, on educational and social functioning is important. There is some evidence of the longer term impact of BECTS on cognition. Monjauze et al. (2011) investigated the performance of young people in remission from BECTS using assessments of IQ and language skills and compared this with age matched controls. Findings demonstrated that despite being in remission, 6 of the 13 participants scored below average on a standardised language assessment,

indicating that specific language impairments may persist beyond the active phase of the disorder [11]. Continuing motor difficulties were again not considered.

DCD is a developmental disorder where the motor impairment experienced is significant enough to interfere with day to day activities and academic achievement [12]. It is known to affect up to 5-6% of the population [13] and motor difficulties have been shown to continue in around 60-70% of individuals [14]. There is extensive evidence to show increased risks for depression and anxiety disorders [15; 16]. DCD has also been shown to co-occur with ADHD, ASD, Dyslexia and Specific Language Impairments [17; 18; 19; 20; 21]. The criteria for diagnosis, as in the DSM-5 (American Psychiatric Association, 2013), requires the use of a standardised measuring of motor impairment along with considering the impact on activities of daily living [12] The most common assessment used in the UK is the Movement ABC-2 Battery [22]. Handwriting difficulties are commonly seen in DCD [23].

The aim of the study was to identify the pattern and level of language, cognition, and in particular a focus on the less studied area of motor functioning in a group of children with BECTS using a battery of standardised measures.

The data reported here were collected as part of a collaborative project between an NHS Trust, a brain imaging research centre and a clinical and research centre specialising in developmental disorders.

2. Method

2.1. Recruitment

Children aged between 8-16 years were identified through general and specialist NHS paediatric clinics in South Wales, UK. The families of children who met the criteria for a BECTS diagnosis as reviewed by two paediatric neurologists (FG and JtWN) were invited to take part in the study. Inclusion criteria were a clinical diagnosis of BECTS supported by EEG showing centro-temporal spikes. After getting a thorough description and understanding of the study all participants gave assent and parents gave informed consent on behalf of the child.

2.2. Participants

Twenty-four children were recruited. Three cases were excluded. One child had Klinefelter's syndrome, one was diagnosed with Polymicrogyria after recruitment, and one child was unable to complete the assessments. In total, 21 children (age range 8 years 1 month-15 years) were entered into the study, which was part of a larger neuroimaging study [24]. The mean age of the group was 10 years 8 months (SD 1.98). Nine of the children (42.9%) were female and twelve (57.1%) were male (1:1.3 ratio).

2.3. Measures

Along with motor measures, a measure of intelligence and language functioning was also sought to consider the level of variability within this group. The following standardised assessments were administered in order to evaluate the type and degree of impairment present in the children with BECTS compared to normative data. All of the measurements derived percentile scores whereby a child's performance is scored from 1 to 100. Because all assessments were standardised on normed groups it was not felt it was necessary to have a control group for this study.

2.3.1. The Kaufman Brief Intelligence Test 2 (KBIT-2) [25]

The KBIT-2 provides a quantitative estimate of intelligence and can be divided into 3 distinct scores: verbal, nonverbal and an IQ composite. This study has used the following as descriptive cut-offs: (a) 16th percentile and above indicates no difficulty, (b) 6th to 15th percentile indicates some moderate difficulty is present and (c) 5th percentile or below indicates a severe difficulty.

2.3.2. The Test for Reception of Grammar 2 (TROG-2) [26]

The electronic or paper version of this was administered to evaluate the child's understanding of grammatical constructs, using a multiple-choice format. The test consists of 80 four-choice items. This study has used the following as cut-offs: (a) 16th percentile and above indicates no difficulty, (b) 6th to 15th percentile indicates some moderate difficulty is present and (c) 5th percentile or below indicates a severe difficulty.

2.3.3. The Movement Assessment Battery for Children 2 (MABC-2) [22]

This is a standardised series of measures used to identify impairment in motor functioning. It was used to gain overall standard and percentile scores for manual dexterity, aiming and catching and balance subsections and a total score. The assessment produces percentile data which can be interpreted as follows: (a) 16th percentile and above indicates no movement difficulty present, (b) 6th to 15th percentile indicates that moderate movement difficulty is present and (c) 5th percentile or below indicates a severe difficulty.

2.3.4. The Developmental Coordination Disorder Questionnaire (DCDQ-'07) [27]

The DCDQ is a parent completed screening tool used to screen for DCD and considers functional impairments. Parents compare their child's motor performance to that of peers according to 15 items. These are grouped into three subsections: control during movement, fine motor/handwriting, and general coordination. The questionnaire produces a cumulative score, which is also categorised as 'suspected DCD' (S) or 'not DCD' (N) for that age.

2.3.5. The Detailed Assessment of Speed of Handwriting (DASH) [28]

The DASH was used to test handwriting speed and quality as difficulties are commonly associated with DCD. The assessment, made up of five subtests, is designed to examine the precision of fine motor skills, the speed of writing the alphabet, the ability to alter speed of performance on two tasks with identical content and a free writing competency task. The DASH produces percentile data which can be interpreted as follows: (a) 16th percentile and above indicates no handwriting difficulty, (b) 6th to 15th percentile indicates that moderate handwriting difficulty is present and (c) 5th percentile or below indicates a severe handwriting difficulty. This test is only suitable for use with children of 9 years and above.

The study was approved by the local NHS research review and ethics committee.

3. RESULTS

See Table 1. This provides clinical information on the case series.

See Table 2. for case series with percentile scores in the assessments and subtests.

The information is presented as a case series rather than as compacted group data, in order to explore and see the actual variability within the clinical group, that may otherwise have been 'lost'. Hence the reason for the table with scores being presented in this way.

3.1. KBIT-2

Out of 21 children who completed the verbal subtest of the KBIT, 2 (9.52 %) scored on the 5th percentile or below in their overall scores, 4 (19.05%) scored between the 6th and 15th percentile, and fifteen (71.43 %) scored above the 15th percentile. Out of 21 children who completed the non-verbal subtest of the KBIT, 1 (4.76%) scored on the 5th percentile or below in their overall scores, 5 (23.81%) scored between the 6th and 15th percentile, and fifteen (71.43 %) scored above the 15th percentile. Overall out of 21 children the IQ composite component of the KBIT, 1 (4.76 %) scored on the 5th percentile or below in their overall scores, 4 (19.05%) scored between the 6th and 15th percentile, and sixteen (76.19 %) scored above the 15th percentile.

3.2. TROG

Out of the 19 children who completed the TROG measure there was some missing data for the TROG on two cases. One (5.26%) scored on the 5th percentile or below, three (15.79%) scored between the 6th and 15th percentile, and fifteen (71.43%) scored above the 15th percentile.

3.3. DCDQ

Of the 20 responses on the DCDQ, eight (40%) resulted in having 'suspected' DCD. Interestingly, three of these were not children that had scored below the 15th percentile on Movement ABC-2 Battery.

3.4. MABC-2

Out of 21 children, five (23.8%) of the children who completed the MABC-2 scored on the 5th percentile or below in their overall scores, 2 (9.5%) scored between the 6th and 15th percentile, and fourteen (66.7%) scored above the 15th percentile. In the sub tests, six children (28.6%) had manual dexterity difficulties, six (28.6%) children had aiming and catching difficulties, and six (28.6%) had balance difficulties (5th or below percentile). Eleven out of 21 children were shown to have difficulties in one or more subtests.

3.5. DASH

Out of the 16 children who completed the DASH, three (18.75%) scored below the 5th percentile, 6 (37.5%) scored between the 6th and 15th percentile, and 7 (43.75%) scored in the 16th and above percentile. There was one incomplete questionnaire submitted. Five participants did not undertake the DASH due to age restrictions (assessment is for 9 years and above).

4. DISCUSSION

The aim of the current study was to investigate the association between BECTS and the pattern and degree of motor impairment present in a clinical cohort of children. Language and cognition were also measured in the group as this has been noted in other studies previously, and it was of interest to consider the level of heterogeneity within the group. Of interest to note, is that of the 21 children who completed the study, only two (9.52%) were found not to have significant difficulties in any of the measures obtained, and 19 of the 21 (90.48%) participants showed some difficulties on at least one or more areas of functioning. When measuring across three areas crudely defined as language, cognitive and motor functioning, 10 (47.62%) participants had difficulties across two areas, and two (9.52%) participants had difficulties across all three of the domains (9.52%).

With specific relationship to motor difficulties, 5 out of 21 (23.8%) fell below the 5th percentile on 'total' Movement ABC-2 score. In addition to this, the subscores are reported. In The European Academy of Childhood Disability (EACD) guidelines for Definition, Diagnosis, Assessment and Intervention of Developmental Coordination Disorder (DCD) states that: "If a child shows particular difficulties on one domain (i.e., performs below the 5th percentile), but performs above the 15th percentile on other domains, the child should be considered to have a domain specific DCD (e.g., fine motor, gross motor)" [29, p.38].

10 out of 21 (47.6%) children on the assessment, Movement ABC-2 Battery had difficulties in one or more domains (i.e. scores below 5th percentile). Additionally, three out of 16 children had significant handwriting difficulties (scores below 5th percentile), with another six having some difficulties. It is not possible to say whether this is DCD or as a secondary consequence of the epilepsy. However, motor impairment was present in nearly half the children and it would seem sensible for it to be considered when taking a history clinically.

The results demonstrate the wide variability and inconsistency in patterns of presentation *between* cases despite all having the same diagnosis of BECTS. This was a key reason to present the data in this manner, as a case series. The high level of motor difficulties noted in this study adds to the evidence base provided by the work of Scabar et al. (2006) [8] more than ten years ago.

On the basis of the findings of this study and previous research it is recommended that screening for *all* developmental disorders should be considered routinely for children diagnosed with BECTS to identify the presence, specific pattern and degree of impairment including asking about motor functioning. This will allow for the child to be supported appropriately and specifically. While factors may operate independently, there may some evidence to act jointly (interaction effect), or mediate relationship between disorders. Further studies need to consider these potential cumulative and interactional effects.

Similarly, it may suggest that routine history taking relating to BECTS symptoms in children presenting with attention, language or motor disorders should also be considered. Vannest et al. (2015) propose, as others have done, that BECTS exists on a spectrum with other pediatric developmental disorders, both presenting with and without epileptic seizures, and predicting potential prognosis may be dependent on viewing this total picture [1]. This study supports the neurodiverse nature of the condition. Apart from the one study undertaken by Scabar et al. (2006) and a more recent study considering

fine motor function by Vannest et al.(2016)[31], few studies have been specifically undertaken in children with DCD examining the potential presence of BECTS, and this remains not routine clinical practice [8]. A careful history of nocturnal or possible seizure like events would seem to be prudent in all children diagnosed with DCD. At present there is insufficient evidence to recommend routine EEG and further research is required to confirm or refute the findings of Scabar et al. (2006) [8]. A longitudinal study alongside serial MRI measures could also potentially help to understand why some children with DCD improve in adolescence [30].

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