**UNIVERSIDADE DA BEIRA INTERIOR**

**FACULDADE DE CIÊNCIAS DE SAÚDE**


# BENIGN CHILDHOOD EPILEPSY WITH CENTRO-TEMPORAL SPIKES

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CORRELATION BETWEEN CLINICAL,

NEUROPSYCOLOGICAL AND

ELECTROENCEPHALOGRAFIC ASPECTS

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***“Symptoms then are in reality, nothing but the cry from suffering organs.”***

Jean Martin Charcot

# DEDICATION

I dedicate my success to my mom and my dad, whose unconditional support made this dream possible.

And to my sister Sandra and brother Daniel, you are my role models and best friends. With you by my side, all things seem possible, thank-you for teaching me to never give up.

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# ABSTRACT

Benign childhood epilepsy with centro-temporal spikes is a form of epilepsy with no demonstrable anatomic lesion showing spontaneous remission. Seizure frequency is low, typically 2-5 total seizures, but also quite variable, ranging from a single lifetime episode to multiple seizures per day. The prognosis is excellent, with 98% of children outgrowing the disorder by puberty. Although the absence of cognitive deficits has always been considered a prerequisite for diagnosis of this illness, recent studies have revealed mild neuropsychological impairments in different areas, including language, attention, executive functions, memory, visuo-spatial orientation, and phonological processing. These cognitive deficits may be associated with learning difficulties and decline in school performance, which longitudinal studies have shown to be transitory in the majority of cases.

The aims of this study were to assess the types of cognitive problems which may be encountered in this population, to evaluate the course of cognitive and learning capacities during the active phase of epilepsy, and to see if there was a correlation with paroxysmal activity on the electroencephalogram.

The study combined retrospective neuropsychological and electroencephalografical results of 8 children presenting with typical benign partial epilepsy with Rolandic spikes and occipital spikes as well as the results of 5 children belonging to a hospital-based control group. In total, 13 children with ages ranging between 5 to 11 years were included and the following tests were carried out: anamnesis, neurological examination, electroencephalogram, and the Wechsler Intelligence Scale for Children or Wechsler Preschool and Primary Scale of Intelligence-Revised.

As a result of this study's various limitations, namely the limited sample size, no significant correlations were found between the factors connected to the epileptic nature of BECTS, such as the number of seizures, age of onset of the disease, CTS lateralization and the results in the neuropsychological tests.

Therefore it remains to be shown whether these correlations exist, as the neurobiology of Rolandic discharges and their relationship to cognitive dysfunction and epilepsy require further study.

## KEY-WORDS

Benign Rolandic Epilepsy of Childhood (BECTS), centro-temporal spikes, neuropsychology, cognition, intelligence quotient, distractibility.

# RESUMO

Epilepsia rolândica benigna da infância é um tipo de epilepsia sem lesão anatómica demonstrada que remete espontaneamente. A frequência das crises geralmente é baixa, tipicamente entre 2-5 crises em todo o percurso da doença, mas é uma doença muito variável, podendo ocorrer desde uma única crise na vida até multiplos episódios por dia. O prognóstico é excelente, com remissão da doença até à puberdade em 98% das crianças. Apesar da ausência de defeitos cognitivos ser considerada um pré-requisito para o diagnóstico da doença, estudos recentes revelaram défices neuropsicológicos ligeiros em várias áreas, incluíndo na linguagem, na atenção, nas funções executivas, na memória, na orientação visuo-espacial, e no processamento fonológico. Estes defeitos cognitivos podem ou não estar associados com dificuldades na aprendizagem e uma queda no desempenho escolar, existindo estudos longitudinais que demonstraram que estes defeitos são transitórios na maioria dos casos.

Os objectivos deste estudo incluíram: determinar os tipos de problemas cognitivos que podem surgir nesta população, avaliar o curso das capacidades cognitivas e de aprendizagem na fase activa da doença, e definir se existe ou não uma correlação com a actividade paroxística no electroencefalograma.

Este estudo comparou retrospectivamente os resultados neuropsicológicos e electroencefalográficos de 8 crianças com epilepsia típica benigna com pontas centrotemporais, com os resultados de 5 crianças constituíndo o grupo controlo. No total, 13 crianças com idades compreendidas entre os 5 e 11 anos foram incluídas no estudo, e subsequentemente fizeram-se as seguintes avaliações: anamnese, exame neurológico, electroencefalograma, e a Escala de Inteligênciade Wechsler para crianças

– Terceira Edição ou a Escala de Inteligência de Weschler para a Idade Pré-Escolar e Primária – Edição Revista.

Tendo em conta as limitações do estudo, não se encontrou correlação entre os aspectos relacionados com a natureza epileptiforme da epilepsia rolândica benigna, como o número de crises, a idade de instalação da doença e a lateralização das ondas centrotemporais com os resultados das avaliações neuropsicológicas realizadas.

Contudo, permanence incerto se estas correlações existem, sendo necessária a realização de estudos futuros nesta área.


## PALAVRAS-CHAVE

Epilepsia Rolândica benigna da infância, Ondas Centrotemporais, Cognição, Neuropsicologia, Quociente de Intelligência, alterações de comportamento.

# INDEX

# INDEX OF TABLES/GRAPHS

## TABLES



## GRAPHS

# ABBREVIATIONS

ADHD - Attention Deficit Hyperactivity Disorder

AED – Anti-epileptic drugs

BECTS – Benign Childhood Epilepsy with Centro-Temporal Spikes

CBZ – Carbamazepine

CHCB – Centro Hospitalar Cova da Beira

CTS – Centro-temporal spikes

CZ and PZ – Midline electrodes which follow the 10-20 system

EEG – Electroencephalogram

IQ – Intelligence quotient

LVT – Levetiracetam

PIQ – Performance Intelligence Quotient

SD – Standard deviation

VIQ – Verbal Intelligence Quotient

VP – Valproic Acid

WISC-III – Wechsler Intelligence Scale for Children

WPPSI-R – Wechsler Preschool and Primary Scale of Intelligence-Revised

# INTRODUCTION

Benign childhood epilepsy with centro-temporal spikes (BECTS) is the most common partial epilepsy syndrome in paediatric patients [1-8]. The syndrome is characterized by (a) the onset of the seizures between 2 and 14 years of age (usually between 3 and 10 years) [9,10], (b) simple partial motor seizures as an exclusive or dominant type of seizure in the vast majority of cases, (c)characteristic EEG foci occurring on a normal background tracing in the lower Rolandic (Sylvian or "mid-temporal") area, and (d) the absence of neurologic or intellectual abnormalities before and during the period of seizure activity [9].

In children with BECTS, the frequency of seizures is usually low [2,8], with 25% of patients having a single episode and 50% having fewer than 5 fits [9]. The Partial motor seizures preferentially involve one side of the face [9,11,12]; the oro-pharyngeal muscles [9,13]; on occasion, the upper limb (mostly consisting of clonic jerks); and in very rare cases, the lower limb [9]. Facial seizures consist of a tonic contraction of one side of the face and/or clonic jerks of the cheek and eyelids [3]. Oro-pharyngeal signs comprise one or several of the following: guttural sounds; movements of the mouth; the contraction of the jaws; a feeling of suffocation and profuse salivation [9,11]. Sensory phenomena sometimes occur as well, most often involving buccal paresthesias involving the corner of the mouth, the inside of one cheek, the tongue and gums, or teeth [9,11]; children retain consciousness throughout the seizure in most attacks and the arrest of speech with preservation of comprehension is also a frequent finding [9,11,13].

Strictly localized seizures are extremely brief, lasting from seconds to minutes; they occur more often in children older than 5 years of age and are often diurnal, especially upon waking [9]. In younger children, seizures tend to be less localized (possibly involving half of the body), are more often nocturnal, and with a longer duration compared to more localized attacks. Some impairment of consciousness is the rule with prolonged fits [9]. At times, generalized seizures may be the only ictal manifestation, but seeing as they are mainly nocturnal, they may easily go unrecognized [9,11,14]. In more than 50% of patients, seizures occur only during sleep, 5-25% occurring exclusively during wakefulness, arising in both states in the remaining children [9,11,14].

The inter-ictal EEG abnormalities of BECTS patients include: a negative sharp wave (>100μV) with a relatively blunted peak, followed by a prominent positive wave whose amplitude may be up to 50% of that of the preceding sharp wave [9,15-17] . No relationship is found between the frequency or extent of the sharp waves and the seizure frequency and duration [9]. The discharges usually appear on a normal background rhythm, and are mainly localized to the centro-temporal (Rolandic) region of the hemisphere contra-lateral to the clinical seizures [18]. However, in younger children they may be located in more posterior regions. In 33% of patients, sharp waves occur bilaterally [9].

The absence of frank inter-ictal neurologic or mental impairment is one of the BECTS criteria [1]. However, several studies argue that severe encephalographic epileptiform activity in childhood causes cognitive deterioration by disturbing the formation of synapses [14]. The reported disturbances include a possible effect on language lateralization on the side opposite the focus; small differences in cognitive

performance, mainly on tests of attention and visuo-motor skills between patients and controls; intellectual and behavioural deficits on a battery of neuropsychological tests and language dysfunction on some tests [4,5,9].

## OBJECTIVES

The significance of these abnormalities is not easy to assess. Therefore, the purpose of this study was to compare the cognitive functions (as assessed by the WISC-III and the WPPSI-R) of BECTS children divided into 2 groups (those with and without epileptic seizures) with children of a hospital-based controls of the same age group.

# MATERIALS AND METHODS

Eight children with ages between 5 and 12 years diagnosed with BECTS according to the International League against Epilepsy classification were included in this study [19]. The children were chosen during the Child Neurology Outpatients Consults and the Electroencephalography Consults carried out by the Cova da Beira Hospital Centre (CHCB), in Covilhã. Children were only included after the parents and/or guardians gave their consent.

The following procedures were carried out on each child in this study:

1. Medical history and examination of the medical file with the clinical and laboratory data.
2. Neurological examination.
3. The Wechsler Intelligence Scale for Children (WISC-III) – a widely used measure of general intelligence for children aged 6-16 years. The WISC-III is organized into three IQ scores (Verbal, Performance, and Full Scale) and further divided into four factorially derived index scores including the Verbal Comprehension, Processing Speed, Freedom from Distractibility and Perceptual Organization. Each of the IQ scores and factor indexes yield standard scores with a mean of 100 and a Standard Deviation of 15. The WPPSI-R – Wechsler Preschool and Primary Scale of Intelligence-Revised which is test equivalent to the WISC-III for younger children, was applied to one child of the age of five.
4. Digital Electroencephalogram – brain electrical activity was recorded using the Grass Telefactor EEG system, with a resolution of 16 bits, 0.3 and 35 Hz filters, analyzing 200 samples per second. Impedance was maintained below 5 kΩ. The

electrodes were placed according to the 10-20 international system and the reference between CZ and PZ. The recording was carried out while the child was resting, in hyperventilation for three minutes and, when possible, during spontaneous sleep.

The hemisphere of the brain in which the epileptic activity took place was established.

The project was approved by the Investigation Committee of CHCB - Covilhã.

*Data analysis* – A descriptive analysis was carried out to determine the relationship between the clinical and electroencephalographic aspects as well as the cognitive aspects obtained using the WISC-III/WPPSI-R.

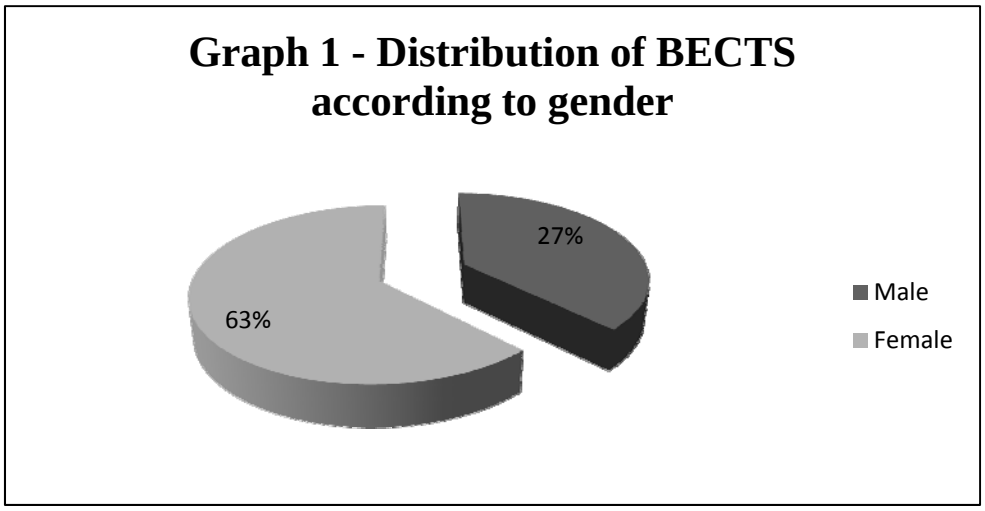
The programs used for statistical analysis included: *Microsoft Office Excel 2007*® and *Confidence Intervals on Probabilities Calculator*® [20].

With the objective of making a comparative analysis of the WISC-III results of the children with BECTS, a hospital-based control group was formed, consisting of 5 children paired with respect to age, presenting no antecedents suggestive of pathologies involving the central nervous system, alterations in the neurological examination or EEG.

# RESULTS

## Clinical aspects

With a study sample of 8 children with BECTS, 5 were of the female gender and 3 of the male gender. Therefore, a female predominance (63%) was clearly evident in this study. In the control group, there were 2 girls (40%) and 3 boys (60%).



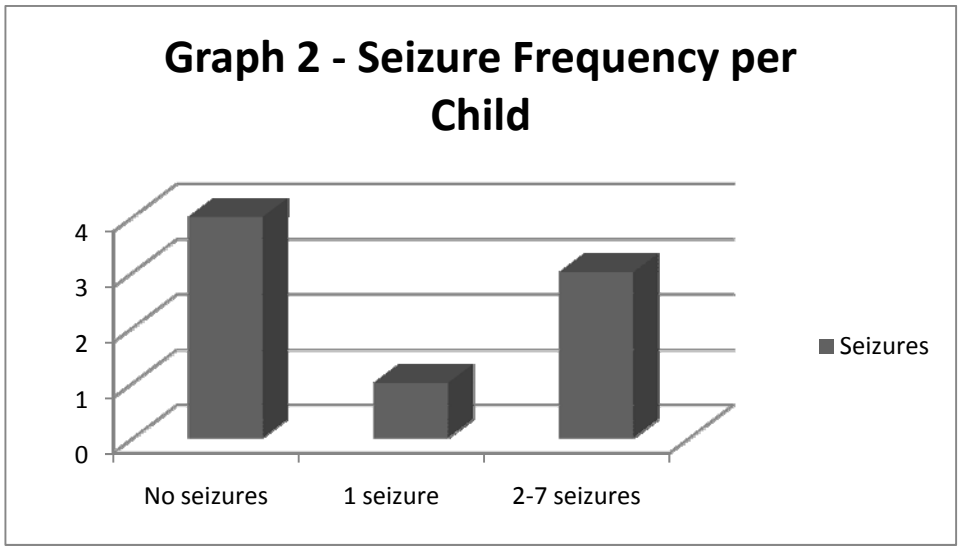

The age of diagnosis for the BECTS children was between 2.5 and 10.4 (Table 2), and all of the children with a prior history of seizures (50%) were prescribed anti-epileptic medication at some point in their disease (valproic acid, carbamazepine, levetiracetam).

One BECTS child (13%) presented a prior history of febrile seizures.

All BECTS children were right-handed, with one left-handed child in the control group (20%).

In the BECTS sample (Graph 2), 4 children presented merely electroencephalographic evidence of epilepsy (50%), one child (13%) experienced only one seizure, and 3 children (37%) had between two and seven seizures. Therefore, the

absence of seizures (50%) was equally as frequent as the appearance of seizures in this study.



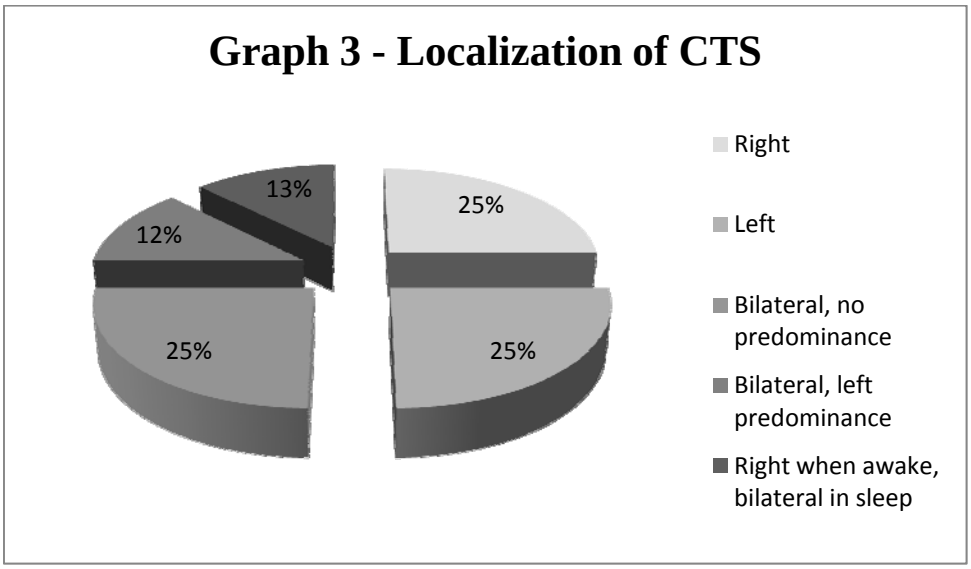

Upon reflection of the data displayed in Table 1, one can conclude that, in this study sample, 94% of the seizures registered were focal, with predominantly oro-pharyngeal (50%) manifestations. Only one seizure (6%) was generalized tonic-clonic, with an oro-pharyngeal manifestation as its initiating focal aspect.

**Table 1 - Frequency of Seizure Type/Manifestations**

| **Seizure type/ Manifestation** | Frequency | Percentage |
|---|---|---|
| **Focal/Oro-pharyngeal** | 8 | 50 |
| **Focal/Motor** | 5 | 31 |
| **Focal/Versive** | 2 | 13 |
| **Generalized/Tonic-clonic** | 1 | 6 |

# Electroencephalographic aspects

At rest, all the children showed normal EEG baseline activity.




Based on the information presented in Graph 3, if we consider the child who displayed CTS in the right hemisphere when awake, and bilaterally in sleep, the majority of the CTS were located bilaterally (50%) although not exclusively. The CTS were exclusively or predominantly in the right hemisphere in 2 cases (25%), and in the left hemisphere in 2 cases (25%).

## Table 2 - Data of BECTS Children

| PN | AD (years) | Seizures (Presence/ Absence) | AED | IQs (full scale, verbal, non-verbal) | Course school/help | Comment |
|---|---|---|---|---|---|---|
| **1** | 8.8 | P | VP | 88 (87/95) | Normal | Attention problems |
| **2** | 10.4 | P | CBZ | 83 (91/82) | Normal | Transient learning difficulties, |
| **3** | 2.5 | P | VP | 69 (72/72)[a] | Day-care, dislalia | Hyperactive, easily distracted, FH |
| **4** | 3 | P | LVT | 125 (142/102) | Normal | Excellent scholastic achievement |
| **5** | 7.7 | A | No | 114 (117/108) | Private | Temperamental, moody |
| **6** | 4 | A | No | 111 (110/111) | Normal | Hyperactive |
| **7** | 6 | A | No | 105 (93/118) | Normal | Attention fluctuations, headaches |
| **8** | 6.5 | A | No | 68 (81/68) | Normal, speech therapy | Enuresis, learning difficulties |

**BECTS: Benign Childhood Epilepsy with Centro-temporal Spikes;**
**PN: patient number, AD: age at diagnosis of epilepsy**
**[a] Difficult cooperation (does not reflect potential); FH, Family history of epilepsy;**
**AED: Antiepileptic drugs; VP: Valproic acid; CBZ: Carbamazepine; LTC: levetiracetam**

## Table 3 - Data of Hospital Controls

| PN | AHC | Chief Complaint | Medication | IQs (full scale, verbal, non-verbal) | Course school/ help | Comment |
|---|---|---|---|---|---|---|
| **9** | 9.7 | Migraines | Topiramate | 96 (101/93) | N | DP, DV, enuresis, HA, FH, |
| **10** | 7.6 | Headaches | Ibuprofen | 74 (82/74) | N, FA | consults in NEPH, PSY, CD |
| **11** | 8.8 | Distractibility | No | 106 (110/102) | N, FA | Very distracted |
| **12** | 9.3 | Panic Attacks | No | 99 (101/98) | N | Also seen by psychologist |
| **13** | 8.7 | Tremor of extremities | Legofer | 86 (87/92) | N | LH, restless, musically inclined |

**PN: patient history, AHC: age at hospital consult, IQ, Intelligence Quotient , N: normal, FA: frequent absenteeism**
**DP: divorced parents, DV: domestic violence, HA: hyperactive, FH Family history, NEPH: nephrology, PSY: psychiatry**
**CD: Child development, LH: left-handed**

## Table 4 - Distribution of children according to WISC-III scores

| | WISC - III | Intelligence Quotient | | | Factor-based Indexes | | |
|---|---|---|---|---|---|---|---|
| | Classification | Verbal | Performance | Full-scale | VC | PO | PS |
| **Cases** | Superior | 1 | 0 | 1 | 2 | 0 | 0 |
| | Average | 6 | 6 | 5 | 5 | 6 | 6 |
| | Inferior | 1 | 2 | 2 | 0 | 1 | 1 |
| **Controls** | Superior | 0 | 0 | 0 | 0 | 0 | 1 |
| | Average | 5 | 4 | 4 | 4 | 5 | 4 |
| | Inferior | 0 | 1 | 1 | 1 | 0 | 0 |

**Superior: very superior and superior; Average: above average, average and below average; Inferior: inferior and limitrophe; VC: verbal comprehension; PO: perceptual organization; FD: freedom from distractibility; PS: processing speed.**

The figures presented in Tables 2 and 4 illustrate that children with BECTS were generally of average intellect (75%), with a low IQ score prevalence of 25% and a 95% confidence interval between 0% and 55% when compared with the control group (Tables 3 and 4), in which the majority (80%) of children had IQ scores within the normal range.

Within the BECTS male population, only 1 child out of three had a low IQ score (34%), while in the female population there was also 1 child out of 5 with a low IQ (20%). In the control group of 3 boys and 2 girls, the only child to score poorly on the full-scale IQ was male (33%). Therefore, females generally did better on the full-scale IQ test than the males, regardless of which group they belonged to.

Regarding the age of diagnosis of BECTS (table 2), participant's 3 and 4 had the earliest onset of seizures, at 2.5 and 3 years of age respectively. However, participant 3 had a very low Full-Scale IQ score and participant 4 had a very high score, making it impossible to distinguish whether the age of epilepsy onset is relevant.

The same can be said for the number of seizures experienced per child. Those children who did not suffer seizures did just as well in the Full-Scale IQ test as the children who suffered from seizures. Participants 3 and 4 had the greatest number of seizures registered, 7 and 6 respectively. Participant 3 had a very low IQ and participant 3 an excellent IQ.

With respect to seizure type, only one child (25%) presented with generalized seizures (participant 1). This child had IQ scores within the normal range in all areas. The remainder of the BECTS children who experienced seizures had merely focal manifestations (75%), and only participant 3 (25%) presented with low scores. Therefore, no association between seizure type and cognitive defects can be made here.

Three children demonstrated significant full-scale IQ discrepancies, with participant 4 presenting a discrepancy of 40 points ($p < 0.01$), participant 7 presenting a discrepancy of 25 points ($p < 0.01$) and participant 8 a discrepancy of 13 points ($p < 0.05$).

When analyzing each IQ sub-test separately, 75% of BECTS children had normal VIQ scores, one child had superior scores, and one child had low scores. In the control group, all 5 children presented normal VIQ scores.

With respect to the PIQ, 75% of BECTS children had normal scores, while 2 children scored poorly. In the control group, the majority of the children had average scores (80%), with only one child having low scores.

In the sub-tests such as verbal comprehension, perceptual organization and processing speed (Table 4), the majority of children had average scores in the BECTS group (100%, 88% and 88% respectively) as well as in the control group (80%, 100% and 100% respectively).

Participant 5 was the only child with a history of prior febrile seizures, yet she had normal scores in all WISC-III sub-tests. Therefore, in this study there appears to be no association between these two aspects.

All four children who experienced seizures were taking anti-epileptic medication. When comparing these children with the non-medicated BECTS children, one child (25) in each group displayed low scores in the WISC-III. Therefore, an association between cognitive defects and anti-epileptic drugs cannot be established.

Upon analysis of the CTS lateralization of each child and their WISC-III scores, those with right CTS (participant's 1 and 6), left CTS (participant's 2 and 7), bilateral CTS with a left-sided predominance (participant 4), and right CTS during wakefulness with bilateral CTS in sleep (participant 4) all presented with normal VIQ, PIQ and Full-scale IQ scores. Participants 3 (low VIQ, PIQ, Full-scale IQ) and 8 (low PIQ and Full-scale IQ) had bilateral CTS with no predominance. Therefore, in this study it can be said that generalized bilateral CTS may have been associated with lower IQ scores.

Six of the BECTS children (75%) also had a history of hyperactivity and/or learning difficulties, in comparison to 2 children (40%) from the control group, as reported by their parents and teachers (tables 2 and 3).

**Table 5 - When to Interpret the Third factor (FD)**

| Factor | PN 1 | | PN 2 | | PN 3 | | PN 4 | | PN 5 | | PN 6 | | PN 7 | | PN 8 | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | FS | SD | FS | SD | FS | SD | FS | SD | FS | SD | FS | SD | FS | SD | FS | SD |
| **VC** | 10 | 2 | 9 | 1 | 10 | 3 | 16 | 3 | 14 | 3 | 11 | 0 | 10 | 0 | 8 | 1 |
| **PO** | 9 | 1 | 8 | 0 | 6 | −1 | 10 | −3 | 12 | 1 | 12 | 1 | 12 | 2 | 5 | −2 |
| **FD** | 6 | −2 | 8 | 0 | 6 | −1 | 14 | 1 | 8 | −3 | 11 | 0 | 8 | −2 | 8 | 1 |

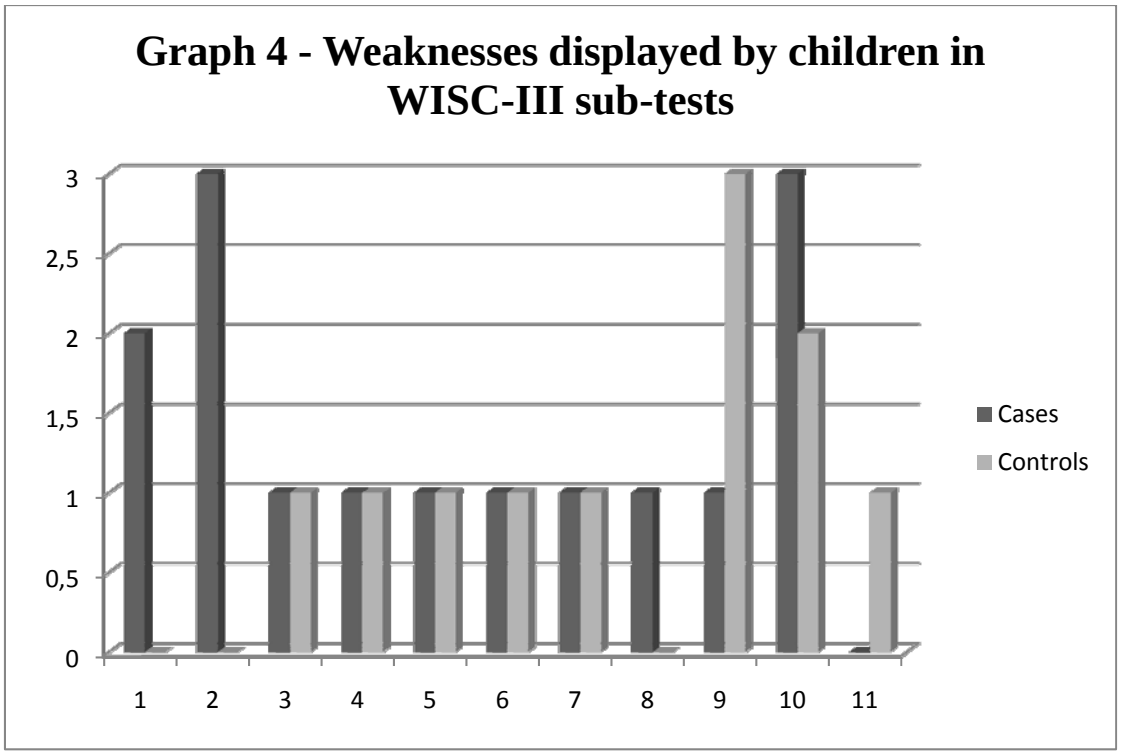
**VC: verbal comprehension, PO: perceptual organization,**
**FD: freedom from distractibility, PN: patient number, FS: factor score,**
**SD: standard deviation from mean**

Based on the results of table 5, three BECTS children (38%) presented WISC-III scores in which the standard deviation from the mean was above or below three, therefore making the Third Factor (Freedom from Distractibility) relevant in these children. In the control group, the third factor was attributable to 2 of the children (40%). Thus, in this study, both groups of children were highly distractible.

Furthermore, based on the WISC-III analysis (table 6, graph 4), several strengths and weaknesses stood out in each BECTS child. Three children (38%) displayed difficulties in arithmetic and mazes, 25% in information, and 13% in vocabulary, comprehension, block design, object assembly, coding, digit span and picture arrangement.

The children belonging to the control group also displayed weaknesses in several areas, including vocabulary (20%), comprehension (20%), digit span (20%), picture arrangement (20%), block design (20%), coding (60%), mazes (40%) and similarities (20%).

When comparing the variety of weaknesses displayed by both groups, it is difficult to draw any conclusions seeing that the BECTS children had difficulties in 5 verbal and 5 performance sub-tests. The children in the control group also had similar difficulties, with 4 verbal and 4 performance sub-test weaknesses. Both groups scored similarly in the mazes' sub-test, and the controls showed extreme difficulties in coding.




1. Information
2. Arithmetic
3. Vocabulary
4. Comprehension
5. Digit Span
6. Picture Arrangement
7. Block Design
8. Object Assembly
9. Coding
10. Mazes
11. Similarities

# Table 6 - Interpretation of VIQ and PIQ discrepancies

| **Subtest** | PN 1 | | | PN 2 | | | PN 3 | | | PN 4 | | | PN 5 | | | PN 6 | | | PN 7 | | | PN8 | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | SS | SD | S/W | SS | SD | S/W | SS | SD | S/W | SS | SD | S/W | SS | SD | S/W | SS | SD | S/W | SS | SD | S/W | SS | SD | S/W |
| **Verbal** | | | | | | | | | | | | | | | | | | | | | | | | |
| Information | 11 | 2 | | 6 | −3 | W | 3 | −3 | W | 15 | −1 | | 13 | 1 | | 11 | 0 | | 8 | −1 | | 11 | 3 | S |
| Similarities | 9 | 0 | | 10 | 1 | | 10 | 5 | S | 19 | 3 | S | 14 | 2 | | 12 | 1 | | 14 | 5 | S | 8 | 0 | |
| Arithmetic | 1 | −8 | W | 9 | 0 | | 6 | 1 | | 19 | 3 | S | 7 | −5 | W | 14 | 3 | S | 4 | −5 | W | 7 | −1 | |
| Vocabulary | 9 | 0 | | 10 | 1 | | 7 | 2 | | 15 | −1 | | 15 | 3 | S | 9 | −2 | | 9 | 0 | | 2 | −6 | W |
| Comprehension | 11 | 2 | | 8 | −2 | | 1 | −4 | W | 14 | −2 | | 15 | 3 | S | 12 | 1 | | 10 | 1 | | 9 | 1 | |
| Digit Span | 10 | 1 | | 10 | 1 | | NA | | | 13 | −3 | W | 9 | −1 | | 10 | −1 | | 9 | 0 | | 8 | 0 | |
| **Performance** | | | | | | | | | | | | | | | | | | | | | | | | |
| Picture Completion | 8 | 1 | | 7 | −2 | | 8 | 2 | | 11 | 1 | | 12 | 1 | | 13 | 1 | | 11 | 1 | | 6 | 1 | |
| Picture Arrangement | 13 | 4 | S | 14 | 5 | S | 5 | −1 | | 12 | 2 | | 8 | −3 | W | 12 | 0 | | 14 | 2 | | 5 | 0 | |
| Block Design | 7 | −2 | | 6 | −3 | W | 4 | −2 | | 10 | 0 | | 13 | 2 | | 13 | 1 | | 14 | 2 | | 4 | −1 | |
| Object Assembly | 11 | 2 | | 8 | −1 | | 3 | −3 | W | 10 | 0 | | 10 | 1 | | 12 | 0 | | 13 | 1 | | 3 | −2 | |
| Coding | 8 | −1 | | 4 | −5 | W | NA | | | 9 | −1 | | 13 | 2 | | 10 | −2 | | 11 | −1 | | 9 | 4 | S |
| Mazes | 6 | −3 | W | 7 | −2 | | 8 | 2 | | 6 | −4 | W | 10 | 1 | | 11 | −1 | | 7 | −5 | W | 5 | 0 | |

**PN: patient number, SS: Sub-test score, SD: Standard deviation from mean, S/W: strength/weakness, NA: not applicable**

# DISCUSSION

It is often difficult to estimate the magnitude of cognitive and educational problems in childhood epilepsy for want of precise definitions and criteria [18]. Several terms such as neuropsychological impairment, cognitive impairment, learning problems, and educational problems have been used in different studies with overlapping meanings.

The classic definition of BECTS requires that there be no neurological or neuropsychological deficits. Prediction of the outcome in individual cases becomes difficult because of the presence of atypical features and minor neuropsychological impairments [18,21,22].

The participants in this study showed clinical and electroencephalographic characteristics of a typical BECTS population, with a low seizure frequency, predominance of focal seizures with sensitive/motor oro-pharyngeal manifestation, an age of onset of seizures between 2 and 10 years, brain activity with normal basal response and no serious behavioural or cognitive complaints [1,4,23].

Data from this small sample of children with BECTS are inconsistent with previous findings of higher incidence in males [2,10], but consistent with data that children are generally of average intellect [1,4,14], with the majority of participants being female and with average IQ scores.

In concordance with some studies [2] and in contrast with others [1], BECTS children in this study presented weaknesses observed in the WISC verbal tasks, such as information, arithmetic, comprehension, vocabulary and digit span. Even so, only one

child (participant 3) scored below average on the WISC Verbal IQ. None of the children in the control group scored below average on the VIQ but they also presented weaknesses in certain verbal sub-tests. Therefore, when comparing the two groups, no specific conclusions can be made.

A reduction in Performance IQ is a frequent finding in BECTS children [24], a fact which was observed in 25% of BECTS children in this study. Participants 3 and 8 presented with Performance IQ scores below those expected for their age, as did participant 10 (20%) of the control group. Following the analysis of these results, it is not possible to determine with any certainty whether these children's scores were influenced by their disease.

Scholastic difficulties in BECTS patients have only recently been recognized and approached, with greater attention being given to specific neuropsychological tests such as language, attention and visuo-spatial perception [25]. However, learning difficulties have been pointed out by teachers and parents [1], with the majority (75%) of BECTS children in this study presenting with histories of transient learning difficulties and attention deficits, with one child receiving speech-therapy.

Participants 10 and 11 of the control group (40%) also received reports from their teachers of inferior scholastic achievement, mainly due to their frequent absenteeism. It is hard to make any assumptions based on this small study, as there are a finite of reasons that cause children to do poorly in school [36], and epilepsy and frequent absenteeism are among them.

Problems in attention and language processing in BECTS children has been hypothesized [26-32], and may be the result of a mild epileptic encephalopathy due to frequent discharges in sleep [14]. Some authors argue that cognitive underachievement

in some children with epilepsy may, in part, be secondary to poor daytime alertness due to sleep fragmentation [14]. Other authors suggest that seizures or subclinical discharges and neuropsychological deficits are different clinical manifestations of the same unknown, genetically determined, pathogenetic mechanism [14].

Symptoms of attention deficit hyperactivity disorder (ADHD) are more common in some particular types of epilepsies, such as Rolandic epilepsy, representing a significant risk factor for academic underachievement [35]. In this study, the Freedom from Distractibility factor, which can have several meanings including (but not limited to), freedom from distraction, ability to manipulate numerical symbols and sequencing ability [36], was applicable to 3 BECTS children (38%) and 2 children from the control group (40%), signifying that they presented with a certain degree of inattention, hyperactivity/impulsivity, or both. Further studies are necessary, however, to reliably confirm a diagnosis of ADHD in these children.

Similar to observations made by many authors [1,4,21], no significant association was observed in the present study between inferior performance in the WISC and the total number and type of epileptic seizure. In addition to this, no significant relationship was observed between inferior performance and the use of anti-epileptic drugs or a prior history of febrile seizures, this being in agreement with various studies on cognition in BECTS children [1].

Although this study sample was too small in number for any significant statistical analysis to be carried out, some worthwhile observations can still be made. For example, participant 3 was the youngest of the BECTS children studied. Being below the age of 5, he was the only child in which the WPPSI-R was performed. Furthermore, this child presented the lowest IQ scores (Full-Scale, Verbal and

Performance), experienced the onset of seizures at the earliest age, and was also the child that experienced the largest number of seizures. The CTS were present bilaterally, and the freedom from distractibility factor was also applicable. However, the neuropsychological evaluation of this child is probably unreliable, as the participant was extremely uncooperative throughout the examination.

On the other hand, participant 4 was also diagnosed at a very young age, experienced only one seizure less than participant 3, also had bilateral CTS, and presented IQ scores within the 95$^{th}$ percentile. Worth noting is that this child´s VIQ (142) and PIQ (102) had a discrepancy of 40 points, which may be predictive of neurological dysfunction. However, VIQ – PIQ discrepancies should not be used to infer neurological dysfunction without convincing support from supplementary data and observations [36], and this child's history points to her being a very bright little girl.

Many studies argue that the later the onset of the seizures, the higher the values observed in the WISC-III [1], but on comparing participants 3 and 4, we become confused as they emerge on opposite sides of the intelligence spectrum, reiterating the fact that more studies need to be done with BECTS patients, with larger samples and over a longer period of time.

The differences between these two cases reinforce the necessity of interpreting IQs in the context of other tests or behaviour-related information before inferring an individual's overall mental functioning [36], as well as indicating the prudence of regrouping subtests in accordance with contemporary practical or theoretical systems to gain insight into the dynamics of a person's cognitive behaviour [36].

Bedoin *et al.* [37] suggest that a relationship exists between cognitive functions and CTS lateralization. Recalling the normal functioning of the brain, one would expect

this be true, with CTS in the left and right hemisphere affecting left and right-brain functioning, respectively.

For most people, the left hemisphere is the primary mediator of verbal functions, including reading and writing, understanding and speaking, verbal ideation, verbal memory, as well as the numerical symbol system [38, 39]. Speaking specifically in terms of the WISC-III, the sub-tests of vocabulary and similarities are specific for left-brain function. However, participants 2 and 7 with left CTS lateralization, scored within normal ranges on these sub-tests.

Right hemisphere language capacities include the comprehension of speech and written material, contribution to the maintenance of context-appropriate and emotionally appropriate verbal behaviour and domination of the processing of information that does not readily lend itself to verbalization [38]. These abilities are evaluated in the WISC picture completion and object assembly sub-tests, in which the participants with right-brain lateralization scored normally.

In this study, only the children with generalized bilateral CTS presented with IQ abnormalities. One child presented with low IQ scores in all 3 IQ sub-sets, while the other had average VIQ scores and low PIQ and Full-Scale IQ scores. Therefore, the results regarding the relationship between Rolandic discharges and higher function impairment are inconclusive.

With respect to CTS lateralization and the Freedom from Distractibility factor, it is less clear whether or not to expect that any correlation exists, being that the prefrontal cortex [38] is one of the main structures involved in attention and that the EEG alterations in BECTS children occur primarily in the central temporal regions of the brain.

However, data from a variety of sources suggest right hemisphere dominance for spatial attention specifically, if not attention in general [38,39]. Other studies suggest that neither hemisphere has an attentional advantage, but rather that each hemisphere directs attention contra-laterally, with the right and left hemisphere directing attention to far space and to near space, respectively.

Furthermore, when interpreting the third factor no suggestion can be made that the child is in any way deficient in his or her numerical problem-solving ability (Arithmetic), short-term memory (Digit Span), or psychomotor speed (Coding) [36]. Such deficiencies may exist, but one cannot infer them from the test administration since the low scaled scores are presumed to reflect the inhibiting effect of a behavioural variable rather than a cognitive or psychomotor deficiency [36].

The child's IQ's must therefore be considered as underestimates of his or her current intellectual functioning because of the behavioural interference, as extreme distractibility or anxiety can easily have some impact on the child's score on any WISC subtest, not just the three associated with the Third factor [36].

This being said, the freedom from distractibility factor was applicable to participants 3 and 4, each of which presented bilateral CTS on their EEG, as well as to participant 5, whose EEG displayed right CTS while awake and bilateral CTS in sleep. No significant correlations can be made in these BECTs children.

In brief, Rolandic epilepsy did not appear to influence the cognitive functions of the children in this study when compared to those belonging to the control group.

Conclusions should be modulated in the light of the small number of patients assessed in this study. However, although BECTS is considered benign with respect to

seizure outcome, it seems important to further explore the qualitative disorders in the developing cognition of children with subclinical epileptic discharges, and the localization of epileptic foci is an important issue.

Whether specific impairments or functional hemispheric lateralization abnormalities persist in adulthood is still undecided [37] and further longitudinal studies are required to answer this clinical question.

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