RESEARCH PAPER

Use of ICF to describe functioning and disability in children with brain tumours

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Abstract

Purpose. To underline the importance of Environmental Factors for reducing Disability and to demonstrate the complex condition of life, especially in terms of social inclusion and participation for children and adolescents affected by brain tumour.

Method. An observational study applying the ICF version for Children and Youth (ICF-CY), the quality of life KIDSCREEN questionnaires and the Vineland assessment was performed. Age-specific ICF-CY Questionnaires were used to interview children at three time-points.

Results. Twenty-nine children were enrolled. Social life and relationships were crucial for defining children’s disability level: formal and informal relationships showed to be very relevant to improve functioning (presence of facilitators in terms of more than 20% of cases). The severity of the disease makes the attention deeply focussed on treatment, neglecting other very important aspects in children’s or adolescents’ life such as their participation in life.

Conclusions. The project highlighted some relevant issues about functioning and disability of these patients, in light of ICF’s Biopsychosocial model of disability. Different rehabilitation projects are necessary for children and adolescents living after brain tumour. Considering treatment and the severity of tumour is very important to define pathways that should also include social and interpersonal aspects.

Keywords: Brain tumour, children with disability, international classification of disease, nervous system disease, public health care, rehabilitation

Introduction

Cancer and acquired brain injury are among the major causes of death and disability [1]. Brain tumours, although less common than the other forms of cancer and brain injury, combines serious health conditions with individuals experiencing associated characteristics of each health condition [2]. For instance, individuals with a brain tumour experience the trauma and uncertain prognosis associated with a cancer diagnosis and the direct neurological effects of the tumour and treatment effects physical, cognitive and behavioural functioning and psychosocial adjustment issues related to change in life circumstances [3]. Many individuals with a brain tumour experience a progressive decline in functioning and a greatly reduced lifespan [4]. As noted by Adelbratt and Strang [5], ‘the disease itself involves a double threat, partly against the life itself and partly against the individual’s personality, owing to the fact that many are stricken with functional and cognitive deficits’ (p. 500). Survival rates among children with a tumour have increased slightly over the past 20 years, with older children evidencing...
better outcomes [6]. Some reports suggest that incidence rates have been rising [7]. The worldwide incidence rate of primary malignant brain and central nervous system tumours is 3.7 per 100,000 persons per year in males and 2.6 per 100,000 persons per year in females. The incidence rates are higher in more developed countries than in developing countries [8]. In children the incidence rate is 3.9 per 100,000 persons per year for 0–19 year olds [9]. Childhood cancer is not a rare event, representing one of the most frequent causes of death in childhood. Childhood cancer is very different from adult cancer as regards type, speed of growth, histopathological characteristics and chemotherapy response. Among children, the most frequent cancer type is leukaemia (33%), followed by nervous system cancer (NSC) (20%), lymphomas (12%), neuroblastomas (8%), soft-tissue sarcomas (7%), renal tumours (6%) and bone tumours (5%). Childhood cancer research represents a real challenge both for general research and for studying its causes. Over the last 20 years, therapeutic results have been more and more consistent, providing a research stimulus in paediatric oncology considering that the benefits in survival and in the quality of life among children are far more evident than the ones obtained in adults [10]. Studies have shown that compromised neuro-behavioural functioning may emerge at any point along the continuum of diagnosis and treatment and those tumour-related sequelae could impact quality of life. In addition, behavioural symptoms including disruption in sleep or eating pattern can predate and may be related to the tumour location. The continuum of psycho-social problems observed in association with brain tumours and their treatments range from general behavioural problems to maladjustment, depressive symptoms and poor self-understanding, to emotional dysfunction and more severe emotional disturbances, including organic personality syndromes and psychosis [11].

In Italy, NSC tumours are the second leading malignancy in paediatric age, corresponding to 19% of malignancies recorded in the AIRTUM database in 1998–2002 in children (age 0–14). The age-standardised incidence rate (IR) was 34.1 cases per million children per year (95% confidential interval, CI: 30.8–37.4). The IR standardised by age on the European population was 34.6 (95% CI: 28.4–40.9). Cumulative incidence up to 14 years of age was 0.52 per thousand (95% CI: 0.49–0.54). The most frequent histological types were astrocytoma (35% of NSC malignancies) and PNET/medulloblastoma (cerebellum) (21%). Twenty-three percent of cases were malignancies of unspecified histological type. Overall, 72.9% of cases were histologically diagnosed, with no Death Certificate Only cases (DCO). NSC cancer was more frequent among boys (IR: 38.6; 95% CI: 33.7–43.5) than girls (IR: 29.4; 95% CI: 25.0–33.8). Incidence rates were similar in the different geographical areas: IR was 32.4 (95% CI: 28.4–36.4) in northern Italian cancer registries; 37.6 (95% CI: 29.7–45.5) in central Italy; 36.4 (95% CI: 27.9–44.9) in southern Italy. Cumulative survival at 5 years from diagnosis for cases incident in 1998–2002 was 62.3% (95% CI: 57.3–67.4%). Survival did not show a significant improvement in the study period from 1988 to 2002. Significant differences are not observed in survival by area of residence, gender or age at diagnosis [10]. Another analysis found that survival for NSC tumours increased first in the Nordic countries and West Germany and then in Western and Southern Europe, the United Kingdom and finally eastern Europe [12–14]. For all cancers combined, 5-year survival increased smoothly over the study period in all regions. The improvement was greatest in the Eastern European countries and, importantly, was a feature of each of these countries individually. The overall improvement may in part be attributed to improved supportive care in the intensive-care setting for acute infections and toxicity related to intensive chemotherapy and also for metabolic complications, life-threatening haemorrhage and other effects of the disease on organ function [15].

In 2006, the Italian Ministry of Health funded a 2-year research project named ‘Cure and Care [C&C] of children with neuro-oncological disease: a pilot study to define multidisciplinary pathways integrated with the International Classification of Functioning, Disability and Health, Children and Youth version (ICF-CY)’. C&C ended in December 2008. The coordination of the C&C Project was done by the Carlo Besta IRCSS Foundation (INNCB) having as partners: the Italian National Institute for Cancer (INT, Milan), the Developmental Neurology Unit of Foundation Besta (INNCB, Milan), the Bambin Gesù Paediatric Hospital (OPBG, Rome) and the Children in Hospital Association-ABIO (Milan). The project aimed to collect data on children and adolescents with brain tumour in the first 6 months after surgery and to measure Functioning and Disability using the ICF-CY Classification [16].

To describe Functioning different ICF-based instruments, such as the ICF-CY Questionnaires [17] and the World Health Organisation Disability Assessment Schedule-WHO DAS II for children or proxy version [18], were used together with the Vineland Adaptive Behavioural Scales, a specific instrument to understand social ability and the level of participation [19] and KIDSCREEN, a health-related quality of life instrument specifically designed for children [20,21].

This article focusses on Functioning and Disability issues and aspects measured with ICF-CY Questionnaires. It especially focusses on the disability
aspects taking into account that the majority of childhood brain tumour research has primarily focussed on motor/sensory functional deficits or on alterations in brain functions or on the neuropsychological deficits in these children [22]. Moreover, epidemiological studies suggest that children with chronic neurological diseases are at significant risk for developing psychological disorders [23]. The study also wanted to evaluate the issue that children with brain tumours may also be at risk for social, emotional, and behavioural problems [24].

Materials and methods

Participants and data collection procedure

Children and adolescents were enrolled in the three clinical units involved in the C&C project. In total, 35 children and adolescents were enrolled.

Eligible participants were children and adolescents from different hospitals who had been diagnosed with a brain tumour – primary and secondary – and who had already had brain surgery, aged between 0 and 18 years. Children and/or adolescents with comorbidity, mental retardation and six children that dropped-out were excluded from the study. The six children were excluded for health reasons and in one case because of death. Patients that were previously enrolled in clinical trials or whose parents were unable to give their consent were also excluded. All recruited patients and/or their parents signed an informed consent form, approved by the Institutes’ Ethical Committees. According to Italian law, adolescents of 13 years and over have the consent form together with their parents. Children and adolescents or their parents or both were interviewed at three time-points (just after surgery T0, after 3 months-T1 and after 6 months-T2) in the three units involved in the project (INNCB, INT, OPBG). The choice to interview the parent, the child or both was taken according to the age of the child and the free decision of the responsible parent.

Measures

The ICF-CY age-specific Questionnaires are a selection of ICF-CY categories divided for age-specific characteristics (0–3, 4–6, 7–12 and 13 – 18 years), and these were used as a basis for data collection. In a questionnaire, a limited number of items from ICF-CY were selected for use based on the judgement that they were considered especially important for a certain age group, developmental stage, context or diagnosis [25,26]. Categories from domains for Body Functions (BF), Body Structures (BS), Activities and Participation (AP) and Environmental Factors (EF) were used at the first and second level. In fact, at this level they are precise enough to serve the need of describing the different issues on functioning and disability at different ages.

Statistical analysis

This study is an observational multicentre trial. Different types of analysis have been done.

For BF, a first level of analysis took into account the relation between the Extension Index (EI) and the Severity Index (SI). The EI corresponds to the number of categories in which qualifiers 1–4 (describing the full range from mild to complete problems) were applied; the SI corresponds to the number of categories in which qualifiers 3 and 4 (describing only severe or complete problems) were applied. After calculating the two variables (extension and severity) at the three time points, we have calculated the mean values of extension and severity for each domain of BF. We show means for all group and each domain. The means of these two indexes were compared through the t-test analysis at the three time-point assessment. For BS, the same analysis of BF was done. The t-test analysis used the Student method with \( z < 0.05 \).

For AP, the same analysis done for BF was done for the two qualifiers performance and capacity. Also in this case for each domain of AP that had problems in our sample, we have calculated four new variables (extension for performance, extension for capacity, severity for performance and severity for capacity). Also the t-test analysis between T0 and T1 and between T1 and T2 means was performed. In addition, to understand the frequency of responses, we calculated the percentage of children and youths that opened codes (had problems) in each domain. With respect to AP, we considered the mean values of the third observation T2.

For EF, a count-based method to analyse data was performed. The number of facilitators, distributed in ‘mild, moderate’ (from 1 to 2), ‘substantial, complete’ (from 3 to 4), counted for each domain. The same counting was performed for barriers.

Results

The final sample of our study was composed of 29 children: 11 males and 18 females (38%, 62%). All age-groups are represented in Table I.

Other children enrolled in the initial phase abandoned after the first or second interview of the study for health conditions, one for death and others for Health problems. These children’s protocols
completed only at T0 or at T0 and T1 were excluded because they were missing the full three observations.

All brain tumours were considered in inclusion criteria and were codified using the ICD-10 Classification [27]. The most represented localisation of brain tumours were in cerebellum (32.4%, ICD-10 code: C71.6), cerebrum (14.7%, ICD 10 code: C71.0) and the parietal lobe (11.8%, ICD-10 code: C71.3).

The geographical distribution of patients shows that the three units enrolled patients is from all over Italian territory (Figure 1).

The majority of interviews were conducted with parents, particularly mothers (65%). All patients were either inpatients at each unit or outpatients, who went to the hospitals for treatment or follow-up. OPBG, Rome, enrolled inpatients and outpatients in day hospital. In the frame of usual collaboration, both being in Milan, INT has specifically sent some inpatients to INNCB to facilitate the enrolment process.

Analysing the ICF CY results showed that all the opened codes were opened in more than 20% of cases. The t-test analysis demonstrates no significant differences in the three-time-point in terms of clinical improvement.

In Figure 2, it is possible to observe which impairments are present and how severe they are. The most important domains that shows significant values are Mental Function (B1), Seeing and Pain Functions (B2), Genitourinary and Reproductive Functions (B6) and Neuromusculoskeletal and Movement-related Functions (B7).

For the BS, the most representative domains were Brain Structures (tumour, s110), Skin and Hair Structures (scar and loss of hair, s810, s840) and Cardiovascular System (central venous catheter, s410).

The most important domains in AP that show significant values are D4 (Mobility) and D8 (Major Life Areas) for Performance. D1 (Learning and applying Knowledge), D2 (General task and demands), D4, D5 (Self-care), D8 for Capacity. Differences between Performance and Capacity, both in Severity and Extension, depend on the role played as Facilitators or Barriers by the EF [Figures 3(a) and 3(b)].

The most important Facilitators are Relationships (formal and informal) (E3) and Attitudes (E4) together with Technologies (E1) and services facilities (E5) (Figure 4).

In this article, we reported only EFs because the presence of barriers is observed only in a few cases (less than 10% of cases).

Table I. Number of children and adolescent distributed by age-group and gender.

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Male (%)</th>
<th>Female (%)</th>
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<tbody>
<tr>
<td>0–3 years</td>
<td>10.3</td>
<td>3.4</td>
</tr>
<tr>
<td>4–6 years</td>
<td>3.4</td>
<td>20.7</td>
</tr>
<tr>
<td>7–12 years</td>
<td>10.3</td>
<td>13.8</td>
</tr>
<tr>
<td>&gt;13 years</td>
<td>13.8</td>
<td>24.1</td>
</tr>
<tr>
<td>Tot.</td>
<td>37.9</td>
<td>62.1</td>
</tr>
</tbody>
</table>

![Figure 1. Sample distributed by geographical areas and hospital.](image1)

![Figure 2. Ratio between severity and extension for BF domains.](image2)
Discussion

All information coming out from C&C project demonstrates that the use of ICF-CY and related instruments are useful in making a working model in the field of paediatric brain tumours. Taking into account the information on Functioning and Disability, and thanks to ICF-CY, the results allow an increase in the understanding the reality of neuro-oncological disease and facilitate the design of a better care pathway that goes beyond just the severity, even if for all periods children were in treatment (chemioterapy, radioterapy, etc.).

The discussion of results first of all focuses on two important points that have to be noticed for a correct comprehension of our data. First, as in many paediatric surveys, parents answered questions in most cases, and the findings and the results are often the proxy report and not directly the children or adolescents’ answers, with all the related consequences. Notably, one of the primary differences in this kind of studies is the source of information (i.e. child, parent, health professionals). The degree of reported problems in the adjustment of children with brain tumours appears to be related to whether the informant is the child himself or an adult caregiver.
Parents report their children to be experiencing more adjustment problems, whereas their children report fewer adjustment problems. Discrepancies between types of informants are a consistent concern in the paediatric psychology literature in general, and with children with cancer in particular [24]. A consistent pattern exists in the literature for more benign adjustment outcomes in self-report studies with higher levels of impairment reported by parents, teachers or staff, especially in this patient population [28]. Specifically, a study reported not only high discrepancies between parents and children’s reports, but also those parents that have a considerable level of specific or general distress and this is relevant information to report [11]. Thus, the results of the studies may reflect difficulties encountered when single sources of information are employed. However, it should also be pointed out that few of the reviewed studies directly compared both parent and child reports of adjustment. In addition, sampling and measurement differences could also account for some of the variability in outcome. At this time, it is difficult to ascertain how accurately these few studies on externalising problems generalise to the population of children with brain tumours. Certainly, controversy exists over the degree to which children with chronic illness, in general, and those with cancer specifically, display externalising behaviour problems [24]. In a review of the literature, Lavigne and Faier-Routman [29] report an increased risk of externalising behaviour problems for children with physical problems and chronic illness; however, they hypothesise that numerous biases may influence such results. For example, they observed that parents are more likely to report both externalising and internalising problems [29], whereas children and their teachers are more likely to report only internalising problems. They further hypothesise that such patterns may be the result of physical limitations that preclude children from displaying classic externalising problems in classroom settings, e.g. being up out of one’s chair. Likewise, they also suggest that teachers may minimise externalising problems because they feel sympathetic toward these children. Thus, such biases may also be reflected in our research on children with brain tumours [24].

The second important point is the condition in which the children were during the period of short-term longitudinal observation. In fact, at that moment of the study, children were still in treatment and they had not yet returned to a usual (according their age and engagements) lifestyle. The presence of chemotherapy and radiotherapy, involving iatrogenic side-effects (lowering of immunity, constipation, general physical weakness, fatigue, hair loss, nausea and vomiting, the presence of ventricular or cardiac catheterisation) should be taken into account in terms of disability instead they are necessary for the healing process.

Another relevant aspect is the psychosocial adjustment which refers broadly to the outcomes associated with the stress of the diagnosis of a chronic or life-threatening condition, such as cancer or brain injury [24]. Given that brain tumour represents a form of both cancer and brain injury, it is relevant to briefly overview perspectives on adjustment to each condition [2].

Long-term outcome studies of non-progressive brain injury have documented persisting physical, cognitive and behavioural impairments which contribute to significant psychosocial dysfunction, including psychological disorders, relationship breakdown, social isolation and loss of productivity [30,31]. A review of theoretical models [e.g. 30–33] identified the following broad influences on psychosocial adjustment and quality of life: (a) pre-injury functioning and circumstances; (b) neurological characteristics or the nature and severity of brain injury and resulting impairments; (c) personal reactions to the injury and recovery process and (d) environmental influences (e.g. social support and rehabilitation opportunities). These aspects are important to consider as part of a biopsychosocial perspective on adjustment and quality of life for people with brain tumour. Other research has shown how children with brain tumours typically differ in a number of fundamental ways from children with non-NSC cancers. Importantly, the risk of cerebral integrity is greater among children with brain tumours, not only because of the critical location of the malignancy and its impact on cerebral tissue, but also because of the iatrogenic effects of neurosurgery, radiation, and chemotherapy. In addition, a decline in cognitive functioning, including cognitive slowing (i.e. decreased processing speed), may subsequently impact on a child’s ability to adapt to the stress of their illness ultimately impacting on their psychological adjustment [34].

Our research with ICF-CY points out some of these aspects, particularly looking at BS and AP domains, and specifically highlighting the importance of Environment in reducing the impact of Disability. In our sample, for the medical problems in Functioning and for all problems related to iatrogenic symptoms, the Environment supports the children totally. These aspects contribute reducing disability coherently with the definition of disability suggested by WHO and other authors [35,36].

All the children and adolescents of our study were strongly supported by parents, healthcare professionals and the general environment, the unique exception is in school activities and related activities (such as school in hospital, going back to school, perform school activities, relate to school mates,
etc.), where they are less supported by the environment and where more work is needed to increase participation once they recover from the brain surgery.

Conclusions

Statistics and data from the literature have shown how the impact of paediatric neuro-oncological tumours represents a relevant problem in public health both for clinical ad social aspects. In fact, the research described in this article has taken into account the problematic of brain tumours and the impact of the iatrogenic effects of treatments not only in relationship with surgery, but especially in terms of Functioning and Disability as defined by the ICF Bio-psychosocial model. This article aimed to underline the importance of environment factors for reducing disability and to demonstrate how complex is the life condition of these children and adolescents, especially in terms of social inclusion and participation.

It is important for healthcare professionals to be aware of the long-term neuropsychological deficits that children with brain tumours often experience, as it is equally important for professionals to be aware of how the stress associated with diagnosis and treatment, as well as the late effects of therapy, can impact on a child’s adjustment and quality of life [24, p. 348]. Family relationships are essential to support children and adolescents and to reduce disability. In fact, familial functioning may also play a role in child adjustment, as children from supportive families show adaptive psychological functioning [Ibidem, p. 376]. This information is consistent with studies that have shown that brain tumour survivors have more difficult patterns of socialisation than people surviving non-NSC cancer, even 2 or 4 years after treatment [11].

Participants of our study viewed the diagnosis as a ‘family disease’ resulting in major changes to relationships and roles and this is consistent with Fox and Lantz [37] who identified the considerable burden experienced by relatives and carers of individuals with brain tumours. Considerable difficulties were experienced in accessing information from within the medical system, particularly with regards to making informed decisions about treatment options.

Cancer survival brings a lot of new challenges for individuals including combined physical, psychological and social difficulties, some of which persist for the rest of their lives [38].

Physically, cancer treatment may damage the heart and endocrine system, impair fertility and sexual functioning [37] and lead to disfiguring bodily changes [39]. Psychologically, many cancer survivors experience a high sense of vulnerability and fear of recurrence of tumours and death [40]. Many cancer survivors also suffer from anxiety, depression and feelings of loss of control [41]. Socially, survivors may face altered or impaired family and social relationships [42].

Similar to other cancers, treatment for a brain tumour may include surgery, radiotherapy or chemotherapy and is often initiated rapidly, leaving little time for patients and relatives to adjust to the diagnosis and to consider treatment options [43–45]. Individuals with good prognosis tumours have to adapt to living with the side effects of the tumour and the treatment received, as well as the threat of recurrence for an extended period of time, whereas those with severe prognosis tumours have a greatly reduced life span [46,47].

In light of all considerations, environment plays a very important role in creating and maintaining pathways that support children and their families in a very complex way and creates opportunities of inclusion and participation. ICF permitted to underline all these aspects and to collect all information about health and disability.

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