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Brain disorders

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Introduction

All mental functioning, be it normal or abnormal, is mediated by the brain. Thus, no child and adolescent psychiatric disorder can be thought of as not being brain related. However, there is a separate category of disorders in which the structure of the brain itself is disordered or in which the basic neurological functions are altered so that normal mental functioning may not result. This is most obvious in those disorders that result from morphological alterations of the brain structure due to a noxious agent or event, or due to a neurobiological deficit that seriously affects the organization and development of the brain.

Classification of brain disorder in childhood and adolescence is not very satisfactory. The major classes of brain disorders as set out in the ICD-10 are derived mainly from manifestations of disorders in adulthood with insufficient consideration of developmental aspects in childhood and adolescence. Thus, in contrast to most of the remaining chapter in this volume both ICD-10 and DSM-IV are not considered as the relevant framework for classification of brain disorders in childhood and adolescence.

In this chapter the following major brain disorders with a basic neurological alteration of brain structures and functions will be described: injury, infectious disorder, cerebral palsy, epilepsy and brain tumours. In an additional section the concept of minor brain dysfunction syndromes will be discussed. This concept has been very influential in the past and has largely been ignored in the more recent academic debate in child and adolescent psychiatry. Given its remaining relevance in practical child and adolescent psychiatry, an attempt will be made to identify those elements in the concept that reflect the notion fruitfully and validly that there is a continuum of brain-related symptoms between neurologically

A Clinician's Handbook of Child and Adolescent Psychiatry, ed. Christopher Gillberg, Richard Harrington and Hans-Christoph Steinhausen. Published by Cambridge University Press.

defined structural brain disorders, and the concept of minor brain dysfunction syndromes.

Given the similarities of psychopathological features and mechanisms in the various brain disorders, some repetition cannot be avoided in the present chapter. However, it was decided to have relatively complete descriptions of the various phenomena and issues of each brain disorder so that the busy clinician will get sufficient information from each section of this chapter.

Brain injury

Definition and classification

Traumatic head and brain injury results from an extended force that insults the brain and leads to a transient or persistent impairment of physical, cognitive, behavioural or emotional functions. It may be divided into open and closed head injuries. Open head injury is defined by penetration of the brain, e.g. a depressed skull fracture with underlying cerebral laceration. Usually localized brain damage is involved. Closed head injury is more common, secondary to traffic accidents or to a fall. The resulting damage may be marked by contusions, intracranical hematomes, intraventricular, subarachnoid, subdural and epidermal hemorrhages and contrecoup injuries opposite to the initial impact. Furthermore, diffuse damage of the brain may be a sequel.

A common classification of the severity in the acute stage distinguishes mild, moderate and severe brain injury. The differentiation usually is based on the extent or duration of coma and the posttraumatic amnestic period. Derived from adult traumatology, the Glasgow Coma Scale (GCS) has also been used most frequently in the population of children and adolescents. The GCS measures eye opening, verbal response and hand or leg movement by rating each response on a scale of 1 to 5 and aggregating all three items. Higher scores represent better responsiveness and prognosis.

According to the GCS mild brain injury is defined by a score of 13 or more, posttraumatic amnesia of less than 12 hours or a loss of consciousness for 5 minutes or less. Moderate brain injury is defined by a GCS score of 9 to 12, a post-traumatic amnestic period of 12 to 24 hours, or a loss of consciousness between 5 and 60 minutes (or even more in some studies). Finally, severe brain injury is associated with a GCS score of less than 9, or post-traumatic amnesia lasting more than 24 hours. However, the GCS has been criticized for being often too crude a measure for children. Comparisons of the research literature are hampered by different definitions of the depth and length of coma as a measure of severity. Several studies in children converge in using the above-mentioned GCS total scores as an indicator

of the depth of coma and extending the length of post-traumatic amnesia to 7–14 days in order to define severe brain injury.

Epidemiology

Incidence and prevalence rates

Traumatic brain injury is a very frequent cause of morbidity and mortality in childhood and adolescence. Estimated incidence rates range from 185 per 100 000 children from infancy to age 14 per annum to 295 per 100 000 adolescents and young adults aged 15 to 24 per annum in the United States. With a rate of 550 per 100 000 and per annum, the risk is highest among the 15–19 year olds. Figures around 45 000 children under 16 years with the number of deaths per annum being around 300 have been established in the United Kingdom. British figures document that 10% of children admitted to accident and emergency departments will have a moderate brain injury, and 1 per cent will have a severe injury. As many as 2.5 per cent of children may have sustained a head injury leading to admittance to accident and emergency departments during childhood.

Sex ratios

In terms of prevalence of brain injury, boys outnumber girls 2:1 with a lower rate in young children up to 5 years of age. The gender discrepancy emerges in infancy and is most prominent during school age and adolescence.

Implications for clinical practice

Head and brain injury is a leading cause of mortality and disability in young people and one of the most common causes of chronic brain syndromes in children. In some cases the injury results in transient or even permanent physical and/or cognitive and/or behavioural and emotional deficits. Highly sophisticated professional skills, including expert knowledge in psychopathology, are needed in order to assist the children who are victims of head and brain injury.

Clinical picture

Main features and symptoms

The symptoms due to brain injury vary considerably depending on cause, severity and type of head injury (open vs. closed), additional pre-morbid functioning and age of the child, post-traumatic coping and quality of the psychosocial environment. The various symptoms may be grouped on three functional levels as shown in Table 1.1.

On the neuropsychiatric level the clinical picture differs according to the phase of the disorder.

Table 1.1. Functional sequelae in brain injury

A. Neuropsychiatric
- Acute symptoms
  - Loss of consciousness, agitation, loss of orientation, short attention span
- Transient symptoms
  - Amnesia, slowing of impulse, affective lability, irritability, hallucinations, thought disorder
- Chronic symptoms
  - Psychopathology: agnosia, apraxia, dementia, disinhibition, hyperactivity, attention deficits, personality changes
  - General psychopathology: emotional and conduct disorders, adjustment disorders

B. Neurologic
- Headaches
- Sleep disturbances
- Epilepsy
- Hydrocephalus
- Spasticity
- Movement disorders
- Apallic syndrome

C. Endocrine
- Hormonal disturbances due to
  - Posterior pituitary deficiencies
  - Anterior pituitary deficiencies
  - Hypothalamic pituitary axis dysfunctions

D. Neurocognitive
- Impaired intellectual functions
- Reduced speed of information processing
- Language and communication skills impairment
- Impaired learning and memory
- Attention deficits
- Perceptual deficits
- Executive functions deficits

E. Educational
- Impaired progress in school
- Failing a grade
- Special education provision
- Scholastic skills deficit

F. Psychosocial
- Dysfunctional individual adaptation
- Dysfunctional family adaptation
- Social disintegration

- In the acute phase varying degrees of depth of coma dependent on the severity of the trauma are noticeable with a loss of consciousness even missing in some children. Mild head injury typically is associated only with transient symptoms of dizziness, headaches, confusion and fatigue with no loss of consciousness or a loss of consciousness not exceeding 20 minutes. A diagnosis of delirium may be stated when agitation, loss of orientation and short attention span are evident.
- There may be a transient phase of relatively short-lived psychopathological features due to a post-traumatic impairment of mental functioning including memory, impulse, affects and thought. Minor transient psychopathological syndromes include slowing of impulse, loss of initiative, forgetfulness, emotional lability and irritability. Moderate to severe transient psychopathological syndromes may include prolonged amnesia and even psychotic symptoms, like formal thought disorder, hallucinations and paranoid symptoms.
- In the chronic phase various specific impairments may become evident. In some children localized deficits leading to the syndromes of agnosia, apraxia and aphasia may result from the injury. A persistent intellectual impairment may justify a diagnosis of childhood dementia and a persistent behavioural pattern resembling a frontal lobe syndrome in adults may be emerging. This syndrome of social disinhibition is marked by a lack of impulse control, hyperactivity and attention deficits and may be accompanied by forgetfulness, talkativeness and carelessness. Owing to the persistent change in the child's behavioural features, a post-traumatic personality change may become noticeable.

  Even more common in the chronic phase are symptoms of general psychopathology. With the well-established increased risk of any child psychiatric disorder in brain disorder, there may be an interaction between neurological and behavioural factors in addition to separate paths for the two factors contributing to specific psychopathologies. These interactions may result in oppositional defiant and conduct disorders, anxiety disorders or affective disorders or rather than creating symptoms *de novo* may amplify a pre-existing hyperkinetic disorder. Among the adjustment disorders the possibility of some symptoms or, less common, the full picture of post-traumatic stress disorder should be considered.

In addition to the various neuropsychiatric features, there is a wide range of symptoms on the neurologic level.

- Various post-traumatic headaches resemble other chronic headaches like migraine and tension headaches, cluster, cervicogenic and myofacial pain headaches. In association with dizziness, nausea or vomiting, other problems of the trauma like hematomas, arterial dissection or aneurysm have to be ruled out.
- A sizeable proportion of patients with head injury experience sleep disturbances. The most frequent early symptom after head injury is hypersomnolence. It is

assumed that hypersomnolence results from damage of the reticular formation or the posterior hypothalamus. Insomnia is less common after brain trauma.

- Post-traumatic seizures at the time of the impact do not increase the risk of epilepsy necessarily. However, this is the case for seizures occurring later in recovery. In contrast to adults, children are more prone to develop early seizures and status epilepticus.
- Another potential complication of head injury is hydrocephalus, especially when there is subarachnoial or intraventicular hemorrhage. Hydrocephalus occurs only rarely when there is massive hemorrhage. The typical manifestation is months later.
- Only in the most severe head trauma does spasticity occur. Resulting from an impairment of corticospinal pathways, fine and gross motor co-ordination and dexterity are affected. Spasticity is marked by an increased tone associated with hyperreflexia.
- Appearing only months to years following the injury movement disorders are a relatively rare complication. Most frequently, the symptoms are due to damage of the basal ganglia or the nigrostriatal pathways leading, for instance, to dystonia of a hemiplegic limb or choreoathetosis. Tremor may follow even mild traumatic brain injury.
- Severe brain trauma may result in rare cases in the apallic syndrome characterized by a functional disconnection of the cortex and the brainstem. The main symptom is the so-called coma vigile with the patient having a markedly reduced consciousness without content and activities while being awake and reduced to a few basal autonomous functions.

Also, the endocrine system may be affected by traumatic head injury in various ways.

- The involvement of the posterior pituitary may lead either to excessive retention of free water or to diabetes insipidus with inappropriate or insufficient secretion of antidiuretic hormones.
- Damage to the anterior pituitary may result in dysfunction of various target organs. Thus, short stature may result from deficiencies in growth hormone, hypothyroidism from a deficit of thyroid-stimulating hormone, hypogonadism from deficiencies in follicle-stimulating and luteinizing hormone, and hypoadrenalism from adrenocorticotropic hormone deficits.
- Excessive appetite and also anorexia may be the consequence of post-traumatic hypothalamic dysfunction, and precocious puberty may result from dysfunction of the hypothalamic–pituitary axis.

Further sequelae of head and brain injury are evident on the neurocognitive level.

- Depending on the severity of damage, there is a varying degree of impaired intellectual functions, with a decrement between performance and verbal scale

scores in standardized tests. Non-verbal functioning is more closely correlated to severity of injury than are verbal scores. The former is more dependent on speech, dexterity and problem-solving skills whereas verbal skills rely on retrieval of information that may be overlearnt. The impairment of the general level of functioning may manifest as a loss of skills, a failure to make any progress or a slower rate of learning.

- Another very frequent neuro-cognitive sequel is reduced speed of information processing affecting various other neuro-cognitive functions, i.e. language, learning and memory, attention and perceptual and motor tasks. Functionally, the affected children may be slower in everyday life, including school and leisure time activities, where fast and complex information processing is needed.
- Problems of naming and with expressive and written language and a deficit in verbal fluency will point to language and communication skills impairment. Sometimes the impact of the trauma may also result in transient mutism. Depending on the age of the child, grammar and syntax may be affected in pre-schoolers whereas higher-level language skills may become deficient in older children and in adolescents. Persistent problems may include a lack of prosody, slowed rate of speech or articulation deficits.
- Further problems stem from impaired learning and memory leading to slowed down knowledge and skills acquisition processes, forgetfulness and absent-mindedness that can be seen both in the classroom and at home. Verbal memory deficits may result in failure to recall instructions, or failure to remember can lead to the necessity of repetitions.
- Frequently encountered problems include attention deficits and distractibility. Children with head injuries have problems with both sustained and focused attention and are easily distracted by extraneous events going on around them. Keeping them on task at school or engaging them in activities for a long time at home is not an easy task.
- Among perceptual deficits visual–perceptual motor skills may be affected soon after the injury and these deficits may persist over longer periods in severe injuries. These functional difficulties may be the result of partial loss of visual fields or double vision in a few children.
- Owing to the high vulnerability of the frontal lobes to traumatic brain injuries, executive functions frequently are disturbed. These functions comprise basic attention and orientation, working memory, meta-representation skills, semantic representation skills and a monitoring system. They involve the capacity for self-determination, self-direction, self-control and regulation. Deficits in these areas impair limitations on the child's capacity to adapt appropriately to changes in environmental demands.

The various neurocognitive deficits lead to various problems on the educational level.

- A large proportion of children with head injuries do not make normal progress in school, fail a grade or need special education provision due to less capable performance at school.
- Educational achievement in these children may be hampered by specific scholastic skills deficits in terms of reading backwardness or sometimes even dyslexia, problems with written language and/or deficits in arithmetic skills with dyscalculia in the most severe cases.

Finally, there are various important consequences on the psychosocial level.

- In each instance of brain injury the child and adolescent has to cope with the impairment resulting from the trauma. Thus, a process of individual adaptation has to start that is based on the individual resources and pre-morbid personality features. Depending on these qualities, there may be both risk and protective factors that either contribute to the heightened risk of general psychopathology in brain disordered patients or buffer against it.
- Family resources are of similar importance. A lack of support from the family may stem from dysfunctional adaptation. Usually families, and most specifically parents, undergo a process of adaptation that starts from an initial shock phase with limited understanding and potential for goal-directed actions. An intermittent phase is then marked by various feelings of anxiety, guilt, anger and blame and finally a relatively stable phase of adaptation is reached. Dysfunctional processes of adaptation may be marked by insufficient care of the patient and/or his siblings, increased family disagreements, partnership problems or even break-up of the family. These problems may be even more pronounced if there is grief and loss for another family member who has died in the accident, a parent or sibling suffering from post-traumatic stress disorder as a consequence of witnessing the injury or an injured parent or sibling.
- Beyond the family, the wider psychosocial environment with relatives, friends and peers reacts to the handicapping condition of the child and exerts an influence on the psychosocial adaptation of the affected child. The reactions may vary between isolation and rejection on the one hand and integration and support on the other. The individual child and his family may both be affected by those either negative or positive processes.

Differential diagnosis and comorbidity

The main question of differential diagnosis in brain injury is whether or not a given symptom or syndrome may be delineated directly or only indirectly from the brain disorder. There is little doubt that very early signs of delirium that can be observed, for instance, in the emergency department, i.e. loss of consciousness,

agitation and loss of orientation are related directly to the impact on the brain. However, in the case of lacking information on history, it may not be clear whether there is a closed head injury or an infectious disorder or an intoxication or another noxious agent exerting an influence on the brain. These various causes will have to be ruled out by careful physical examination including laboratory assessments and neuroimaging.

Similarly, transient psychotic features may pose the question whether or not symptoms of schizophrenia may also explain the clinical picture. Owing to the similarity of the main features in the two conditions, differentiation may be difficult in the absence of clear anamnestic data. Most commonly, the transient psychotic features in brain disorders are marked by hallucinations that are closer to reality and less bizarre and strange and by more concrete and trivial paranoid symptoms.

In the absence of a clear history and/or the exclusion of other aetiologies the localized symptoms of agnosia, aphasia and apraxia also deserve careful examinations assisted by neuroimaging in order to rule out causes other than injury, i.e. brain tumours or anomalies of brain vessels.

With the dominant manifestations of general psychopathology, there is less of a question of differential diagnosis or co-morbidity than an attempt to differentiate the various factors that may have contributed both separately and in interaction with the brain injury to the clinical symptoms.

Various sequelae of the brain injury on the neurologic and endocrine level that may lead to the development of orthopaedic disorders (e.g. scoliosis, contractures, arthritis) may be viewed as co-morbid disorders.

Diagnostic instruments and assessment

The main source of diagnosis in brain injury is clinical examination by the use of history taking, observation and neurological assessment. Neuroimaging by the use of computed tomography (CT), magnetic resonance imaging (MRI) and electroencephalogram (EEG) is mandatory in order to get a full and comprehensive picture of the brain injury.

Neuropsychiatric examination of acute symptoms in brain injury will take place only in the emergency room and transient symptoms will also be noticeable most probably in intensive care units, where examination has to be performed in collaboration with neuropeadiatricians and neurosurgeons. Continuous and repeated observations within relatively short intervals of time are needed in order to examine the fluctuating course of symptoms.

The main domain of neuropsychiatric assessment are the chronic sequelae of brain injury that have to be followed over years in some cases with a very protracted course. The majority of symptoms are represented by neuropsychological deficits in the areas of intellectual functioning, speed of information processing, language and

Table 1.2. Areas of assessment, interview questions, and neuropsychological tests (adapted from Middleto

Area of assessment	Interview questions
1. Intellectual functioning	Loss of skills
	Failure to make any progress
	Slower rate of learning
2. Speed of information processing	Time to carry out everyday living tasks
Motor speed	(e.g. getting dressed)
Thinking speed	Grasp what is going on around them
	(e.g. in general conversation)
	Process information in unpressed situations
	Gather and express thoughts
	Respond to questions or requests
	Carry out motor tasks
3. Language and communication skills	Ability to initiate conversations
Expressive language	Clarity of speech and intonation
Receptive language	Word finding
Word finding	Grammatical structure of speech
Written language	Ability to express ideas
	Ability and flexibility to follow general conversation
	Understanding of jokes, ambiguity, and abstract concepts
	Writing, reading and spelling
4. Learning and memory	Forgetting simple instructions
Visual and verbal memory	Losing belongings
Immediate and delayed memory	Forgetting homework
Recall and recognition	Forgetting plans for the next day
	Forgetting what happened yesterday
	Forgetting where things have to be put down in the home
	Need for many repetitions