Psychiatric Disorders in Children and Adolescents with Intellectual Disability

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23.1 Definition, Classification, and Target Symptoms

In the International Classification of Diseases, 10th revision (**ICD-10**), the term correspondent to intellectual disability still is "mental retardation," and it is specified according to current severity on the basis of IQ scores: F70 mild, F71 moderate, F72 severe, and F73 profound mental retardation (World Health Organization 1996). According to DSM-5 commentary in ICD-11, the diagnostic term will be "intellectual developmental disorders".

In the Diagnostic and Statistical Manual of Mental Disorders, 5th edition (**DSM-5**), the diagnostic term is "intellectual disability" (code 319: assigned regardless of the severity specifier), and the various levels of severity (mild, moderate, severe, profound) are defined on the basis of adaptive functioning (based on the categories "conceptional domain," "social domain," "practical domain") and not on the basis of IQ scores (American Psychiatric Association 2013). The diagnostic criteria include deficits in intellectual functioning and in adaptive functioning both with onset during the developmental period.

The approach of the World Health Organization's International Classification of Functioning, Disability and Health (ICF, World Health Organization 2001) is the attempt to integrate medical and psychosocial features of disability and ability in activities in specific environmental and personal context.

M. Gerlach et al. (eds.), *Psychiatric Drugs in Children and Adolescents*,
DOI 10.1007/978-3-7091-1501-5_23, © Springer-Verlag Wien 2014

Intellectual disability is not per se a psychiatric disorder. But the probability of developing a psychiatric disorder is significantly high in patients with intellectual disability (Arron et al 2011; Bouras 2013; Bouras and Holt 2007; Buckles et al. 2013; de Ruiter et al. 2007; Emerson et al. 2010; Forster et al. 2011; Myrbakk and von Tetzchner 2008). Individuals with intellectual disability and co-occurring psychiatric disorders are at **high risk of problems in social adjustment** and at risk of **suicidal behavior**.

Prophylactic pharmacological measures exist only for some disorders that cause intellectual deficits, such as immunization against infections that can damage the brain. This category also includes dietetic measures, such as the phenylalanine-free diet for those suffering phenylketonuria, and hormonal substitution by timely administration of L-thyroxine during the first 4 weeks of life in congenital hypothyroidism. If the intellectual disability is already manifest, however, psychopharmacological therapy is not directed at these static losses but rather at the psychopathological symptoms or psychiatric disorders of the patients with intellectual deficiency. The **indications** for **pharmacological therapy** are therefore essentially the same as for people without mental handicaps. Medication is, as always, only one part of a care program that includes educational, psychotherapeutic, and socio-integrative measures (for review Sturmey 2012).

The prevalence of psychopharmacological treatment is very high in institutions for the handicapped (Robertson et al. 2000). There are several reasons for this, including:

- Psychotherapy for people with intellectual deficiency is still only very inadequately developed, and the necessary framework and therapeutic qualifications are often lacking.
- Diagnostic difficulties. The severity of psychiatric disturbances and uncertain responses to treatment in this vulnerable population may lead to a greater use of psychopharmacological interventions than in youths without intellectual disability.

Cave! Diagnosis is extremely complex (e.g., see Attiah and Antonacci 2008). As the severity of intellectual disability increases, it becomes increasingly difficult to determine if the symptoms are part of a psychiatric disorder (e.g., mutism in depression) or part of the basic limitations of the intellectual development, e.g., the basic limitations in the use of language that prevent the patient from describing his/her psychiatric symptoms.

23.2 Therapeutic Framework

Therapy with psychopharmacological agents is limited to psychiatric disorders included in the ICD-10 and in the DSM-5, and severe psychopathological symptoms that endanger the patient or others (e.g., self-harm, suicidal tendencies, severe aggressive impulse control disorders), but can also be undertaken in order to allow access to pedagogic and psychotherapeutic measures.

A number of **specific concerns** arise during pharmacotherapy of children and adolescents with intellectual disability.

- The **assessment** of the **benefits** and adverse drug reactions (ADRs) of a medication is, for the same reason, **complicated**.
- The **cause** of the intellectual disability is often **unknown** so that an etiology-based treatment is not possible.
- Pharmacological treatment must be undertaken according to the observable behavioral symptoms and the context in which they are presented (i.e., on the basis of behavioral analysis). The more impeded the capacity of patient with an intellectual deficiency to communicate, the greater the necessity to observe such guidelines.
- Psychiatric and other organic comorbidity (e.g., epilepsy, metabolic disease, cerebral

paralysis, blindness, deafness) are more frequently encountered as the severity of mental handicap increases (Einfeld et al 2011), so that the patient may be subjected to multiple treatments, appreciably increasing the difficulty of monitoring the medication's therapeutic drug benefits, its ADRs, and the interactions between different medications being used to treat the patient.

- In patients with organic brain injuries the expected responses to pharmacological agents might not occur; **unusual** and even paradoxical **effects** may result from the unusual cerebral vulnerability in these patients (Barron and Sandman 1985; Handen et al. 1991, 1992, 1994; Kalachnik et al. 2002; King 2007; Matson and Mahan 2010).
- **Compliance** is more **difficult to achieve**, as with increasing severity of the intellectual deficiency, the ability to communicate and autonomous behavioral monitoring are both reduced; assessment of compliance and drug effects must be undertaken by caregivers.
- The decision to initiate pharmacotherapy should be examined with particular circumspection if it is primarily justified as providing relief for overburdened caregivers. Pharmacological therapy should not be employed to compensate deficiencies in institutions for the occupants with intellectual disability.

23.3 Choice of Pharmacotherapy

There have been very few clinical studies concerning the treatment of psychiatric disorders in youths with intellectual disability. As a result, recommendations are consensus-based and have essentially been derived from the empirical data of a number of therapeutic trials (Bramble 2007; Calles 2008; Häßler and Reis 2010; Handen and Gilchrist 2006; King 2007; Matson and Neal 2009; Matson and Hess 2011; Matson et al. 2000; Reis and Aman 1998; Robertson et al. 2000; Shapiro and Accardo 2010; Sturmey 2012).

23.4 Treatment Strategies

23.4.1 General Aspects of Treatment

Therapy with psychopharmacological agents in children and adolescents with intellectual deficiency, on the basis of these considerations alone, must be managed with particular caution. The complexity of the decision to initiate such therapy is often exacerbated by the fact that the patient is often intellectually incapable of granting legal consent. The following **treatment guidelines** have universal validity but should be particularly heeded in patients with intellectual developmental disorder and the comorbidity of psychiatric disorder:

- Before initiating any treatment, the **diagnosis** and the **assessment of the success** of previously employed therapeutic approaches must be considered. The consensus-based treatment recommendations given by Schur et al. (2003), Jensen et al. (2004), and Pappadopulos et al. (2003) are a useful general guideline for the use of antipsychotics but also for any kind of psychopharmacological treatment (see also Fig. 9.1).
- Do not be misled by the impression evoked by a crisis into a hasty decision to initiate a pharmacological intervention.
- Keep the legal framework for your action in mind.
- Have regard for the wishes of your patient, and inform she or her as far as possible about the measures you adopt.
- Have regard of the opinion and decisions of the "legal adult responsible" person who can provide protections for the intellectually deficient patient. This "**guardian**" may sign consent for psychopharmacological treatment and be called upon to give his agreement to each new change in therapy. Having a specifically assigned family member is better than just picking out any family member transporting the patient to the clinic. This person might change with every clinic visit and not be well informed. People with intellectual disability

often react in a vulnerable fashion to centrally active pharmaceuticals.
- Therapy with psychopharmacological agents alone is rarely effective.
- Therapy with psychopharmacological agents must be integrated into an individual multidimensional therapeutic concept.
- Describe the goals of the treatment or the target symptom as precisely as possible.
- The effect with regard to the defined treatment goals must be systematically documented.
- Prolonged prescription of psychopharmacological agents must be subject to critical review.
- Do not withdraw too rapidly a medication that has been employed for longer periods. Reduction of the dosage of anticholinergic pharmaceuticals is often initially associated with a cholinergic imbalance and thus possibly with agitation and irritability.
- Keep in mind the psychiatric ADRs that can occur. Symptomatic deterioration during adjustment of dosage is typical for such ADRs.
- The general principles of therapy with psychopharmacological agents should be applied (compliance, pharmacodynamics, pharmacokinetics, etc.).
- Pharmacological therapy in children and adolescents with intellectual deficiency additionally requires that the caregivers assume responsibility for reliable dosage, administration, and monitoring of effects.

23.4.2 Therapy in Crisis and Emergency Situations

The ideal of pharmacological therapy is based upon diagnosis, behavioral analysis, a psychoeducative framework (explanation of the diagnosis, of therapeutic alternatives, of the nature and use of the medication, etc.), and obtaining consent from the patient and/or the caregiver to employ a drug, preferably indicated for the intended purpose.

In an emergency situation the baseline conditions are quite different. The patient is often acutely agitated, helpless, and extremely vulnerable; the risk of harm to the patient or to others is significant; personnel are exhausted; the patient lacks insight into his disorder; and the relatives expect relief as quickly as possible. There is insufficient time and opportunity for a complete diagnostic evaluation, and obtaining consent may be impractical.

> **Cave**! In such **emergency situations** there is not only a strong temptation to fast-track the initiation of pharmacological therapy, but it is often also necessary to do so. Documentation of such cases must therefore specifically stipulate that it involves acute pharmacological treatment requiring short-term review, initially on a continuous basis, with regard to indication, effect, and ADRs, and can provisionally be regarded only as a transitional therapy.

Table 24.1 provides an overview of the acute pharmacological treatment of psychiatric emergencies (see Chap. 24). Medications that are frequently employed in emergency situations include:
- Anxiolytics, such as lorazepam (calming)
- Sedatives, such as diazepam (sedative)
- Antipsychotics, such as risperidone, olanzapine, pipamperone, melperone, and haloperidol (calm and sedate psychomotor activity)

23.4.3 Most Common Adverse Drug Reactions

The restricted ability of patients with intellectual deficiencies to communicate, their treatment with several different pharmaceuticals, and, in some cases, their altered cerebral responsiveness necessitate that increased vigilance for ADRs be used when treating children and adolescents with intellectual disabilities. The implementation of therapeutic drug monitoring (TDM) is strongly recommended (see Sect. 2.3). Among the **most frequently** encountered ADRs are the following:
- **The anticholinergic syndrome** associated with the use of low-potency antipsychotics and tricyclic antidepressants is a concern. CNS symptoms include agitation, motor restlessness, dysarthria, disorientation, hallucinations,

and cerebral seizures; peripheral symptoms include obstipation, urine retention, fever, mydriasis, and tachycardia.

- **Constipation**. If overlooked in patients with severe mental handicaps, complaints can lead to uncharacteristic behavioral patterns, such as headache, agitation, depression, and sleep disturbances. Obstipation occurs mostly in association with low-potency antipsychotics, tricyclic antidepressants, benzodiazepines, and carbamazepine.
- **Extrapyramidal-motor disturbances**. Akathisia, stereotypic gaze behavior, and parkinsonian psychomotor symptoms may be mistakenly interpreted as expressions of severe mental disability.
- **Hyperactivity, restlessness, and aggressive agitation** are particularly associated with antiepileptic agents, benzodiazepines, and SSRIs.

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