HABILITATIONAL TREATMENT OF A CHILD WITH FETAL ALCOHOL SYNDROME: CASE REPORT

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SUMMARY – Fetal alcohol syndrome is defined by a triad of symptoms such as facial dysmorphology, prenatal and postnatal growth deficiency, and central nervous system dysfunction. It is the result of teratogenic effects of alcohol consumption in pregnancy. The prevalence of fetal alcohol syndrome is 1 to 3 per 1000 live births. From the neurological point of view, there is a possibility of the central nervous system dysfunction. Structural disjunctions are the consequences of fine and gross motor dysfunction, oculomotor dysfunction, and difficulties in sensorimotor integration. From the functional point view, there are complex cognitive disorders and behavioral disorders, attention disorders and impulse control disorders, learning difficulties, and social communication and perception difficulties.

This paper presents a case study of a boy diagnosed with fetal alcohol syndrome at the age of four, monitored by a team of experts including a physiatrist and neuropediatrician. The boy is also included in polyvalent habilitation treatment provided by a speech therapist, rehabilitator and psychologist.

Key words: Fetal alcohol spectrum disorders; Rehabilitation; Caregivers – education; Professional-family relations

Introduction

Fetal alcohol syndrome (FAS) is defined as a group of symptoms consisting of characteristic facial dysmorphology, prenatal and postnatal growth deficiency, and central nervous system (CNS) dysfunction. The characteristic facial dysmorphology includes short palpebral fissures, maxillary hypoplasia, epicanthal folds, thin upper lip, flattened philtrum, hypoplasia of the nasal bridge, and micrognathia (Fig. 1). Some longitudinal studies have shown that some of the craniofacial malformations that are characteristics of FAS may become less visible over time. Growth deficiency starts prenatally, and the weight and length or height of a person with FAS are below the 10th percentile, but they may normalize by adolescence.

According to Gerberding et al., some of the diagnostic criteria for FAS are functional CNS abnormalities. Performance is substantially below that expected for the individual’s age, schooling, or circumstances, as indicated by global cognitive or intellectual deficits representing multiple domains of deficit (or significant developmental delay in younger children) with performance below the 3rd percentile (2 standard deviations below the mean for standardized testing) or functional deficits below the 16th percentile (1 standard deviation below the mean for standardized testing) in at least three of the following domains: (a) cognitive
or developmental deficits or discrepancies; (b) executive functioning deficits; (c) motor functioning delays; (d) problems with attention or hyperactivity; (e) social skills; and (f) other, such as sensory problems, pragmatic language problems, memory deficits, etc. These two ways (global and functional) of meeting the criteria for a functional CNS abnormality have been adopted because of the composite nature of the cognitive/intellectual and developmental measures. Several specific domains need to be assessed individually to determine that multiple functional domains have been affected. It is important to note that global deficits or delays can leave the child scoring in the normal range of development, but below what is expected for his or her environment and background.

Central nervous system is significantly affected with structural and functional alterations including reduction of the cerebrum shape and size, practically in basal ganglia, corpus callosum, cerebellum and hippocampus. The pattern of structural and functional abnormalities varies, depending on how the alcohol exposure coincides with the critical periods of development. Significant abnormalities are detected as disruption in white matter in the perisylvian cortex, frontal and prefrontal cortex, and white matter hypoplasia was found to be more significant than gray matter hypoplasia. There are significant structural and size changes, especially in the anterior and posterior parts of the corpus callosum. Changes in the isthmus splenium are observed and associated with deficits in verbal learning. The cerebellum structure is disrupted in the anterior position and there is a reduced structure of the vermis, which is associated with motor deficits (balance and coordination), and learning.

Fetal alcohol syndrome is a more frequent cause of mental retardation than the two common syndromes (Down syndrome and spina bifida) together. However, according to the literature, only 25% of people with FAS have mental retardation (global intellectual quotient (IQ) less than 70). The level of cognitive abilities can also vary from profound mental retardation (global IQ less than 20) to above the average cognitive functioning (IQ > 120). In different studies in a population with FAS, the average cognitive functioning varies from mild mental retardation to broad limits of normal cognitive abilities (borderline category for age). Deficits are present in all areas of functioning, with progressive deviation associated with verbal functioning and academic skills (reading and writing skills, mathematical skills) in relation to chronologic age.

The prevalence of FAS is from 1 to 7 children per 1000 newborns, while the prevalence of fetal alcohol disorders spectrum (FADS) is as high as 10 to 40 per 1000 newborns. Estimate of the prevalence of FAS among the chronically alcoholic women is 25 per 1000 newborns. However, many studies point to the fact that a large number of children with FAS are not recognized and not diagnosed, and therefore not included in the appropriate early intervention programs. Early diagnosis and involvement of children in the appropriate interdisciplinary habilitation programs will reduce the long-term negative effects of FAS and improve their quality of life.

Case Report

A 4-year-old boy was monitored from the age of 2 months at the Department of Pediatric Rehabilitation, University Department of Rheumatology, Physical Medicine and Rehabilitation, Sestre milosrdnice University Hospital Center in Zagreb, and from the age of 2 years at the Department of Pediatrics, Zagreb University Children’s Hospital by a team of experts (physiatrist, rehabilitator, speech therapist and
psychologist). At the age of 2 months, the boy was included in the physiotherapy program, at the age of three years he was included in the intensive speech and language therapy, and at the age of four years he was included in the intensive special education program. Difficulties in social and emotional development have been continuously present and family relations are unbalanced and complicated. The boy has a legal caregiver.

He was born after his mother’s second pregnancy that was severely influenced by hypertension at 35 weeks of gestation. Delivery was induced by rupture of fetal membranes and completed by cesarean section due to the presence of meconium in the amniotic fluid. His birth length was 47 cm, birth weight 1570 g, and APGAR 5/8. On initial examination, muscular hypotrophy was detected. The neurological status was without detectable aberration. The initial cranial ultrasound showed periventricular hypoxia and post intracerebral hemorrhage grade II. Heart ultrasound showed open foramen ovale. Initial treatment was introduced because of suspected respiratory insufficiency. In the early development, there was an apneic crisis, while hyperbilirubinemia was treated with phototherapy.

The boy received parenteral nutrition with gradual introduction of enteral feeding up to feeding exclusively through a probe. In the first two weeks of life, the boy was very distressed with the clinical signs of abstinence syndrome, but still without confirmation of reliable history. Intense manipulation tremor and increased sweating were observed. Control cranial ultrasound (at the age of 3 months) showed slight asymmetry of lateral ventricles, slightly indented cornu anterius ventriculi lateralis, and the extremely uneven contour of lateral ventricles. The boy has been continuously controlled by gastroenterologists because of eating disorders.

During the first 12 months (body weight 5.7 kg, body length 71 cm), observation confirmed leg flexion in the hips and knees, with occasional leg crossing, the mouth-leg coordination was developed. The boy could sit by himself in the irregular quadrupled position. Stereotyped torso movements and muscular hypotonia with nonspecific hand movements were present. Spontaneous interest in the offered didactic material was reduced, he accepted sensorimotor stimulation. Emotional communication was superficial, and vocal play was not present.

In the second year of life (body weight 6.5 kg, body height 76 cm), the boy was placing by himself in the sitting position with his back bent, the rear defense was absent, while the ability of crawling was insufficient. Verticalization revealed a paraparetic pattern. Medial body line was exceeded by hands, the elbows were in semiflexion, and there was absence of tremor in action. Social contact was inadequate, fixation of objects in the environment and eye-tracking were developed. He started showing interest in the environment, some receptive vocabulary and gesture imitation were developing. Vocal production was at the stage of reduplicated canonical babbling.

In the third year of life (body weight 8.8 kg, body height 88 cm), neuromotor deviation of atactic pattern with stronger involvement of lower limbs, left eye strabismus and left lateral abducens nerve paresis were confirmed by neuropediatrician. The neuropediatrician stated that the extended deviations were greater than it would be expected based on the available data on the prenatal cranial injury and the underlying diagnosis of FAS. He made first steps at the age of 2 years and 6 months. There were clinical signs of paraparesis with feet deformation. Walking was possible with the assistance and in the presence of leg circumduction. Independent activities were without structural elements with sensory exploration. Vocal expressions were in the stage of non-reduplicated canonical babbling. In verbal production, the ability of imitation of animal sounds was developed, and the use of gesture in communication was appropriate. On comprehensive psychological testing with the Razvojni test Čuturić (RTČ) – Scale of Psychomotor Development11, moderate psychomotor retardation was confirmed.

Between the age of 3 and 4 years (body weight 11.3 kg, body height 91 cm), stereotyped movements of the body and the head became occasional, walking was by the paraparesis pattern on a wider leg basis associated with hand movements. Functional game playing was developed, and nonspecific finger movements were more expressive in the presence of tremor. Expressive language development was in the two-word stage and language comprehension had progressed to the stage of understanding and execution of simple orders. Speech mimicking model was observed. According to
the RTČ scale of psychomotor development, at the age of 3 years and 9 months he achieved results that indicated mild mental retardation (IQ 53).

At the age of 4 years (body weight 13.5 kg, body height 106 cm), the boy had an insufficient response on coordination tests, he could walk the stairs with observation. Running was not possible. Attention deficits were present on task performing. The understanding of spatial relationships was resolved. Lateralization of the body was developed with right-hand dominance during manipulations. According to the Peabody Picture Vocabulary Test (PPVT-III-HR)₁², he achieved scores in the range of the 12th percentile (age equivalent of 3 years and 3 months). In spontaneous verbal expression, the boy was using simple, grammatically correct sentences with adequate word naming. There was a gap in the general knowledge. According to the RTČ scale of psychomotor development, at the age of 4 years and 6 months he achieved results consistent with partial mild mental retardation (partial IQ 60).

Discussion

The patient presented continuous slow psychomotor development and growth with disharmony of skill development (gross motor skills, fine motor skills, cognitive skills, verbal skills, nonverbal and social skills). Disproportion in neurological status and clinical signs was confirmed by neuropsychiatrician at the age of two. The CNS impairment was not diagnosed before the age of two, when neuromotor impairment in the form of the left abducens nerve paraparesis and paresis was confirmed. A gap in the neurological signs, according to the literature, is a typical finding in FAS³.

Early physiotherapy intervention (at the age of two months) stimulated development of gross motor skills and motor skills were continuously improved. With continuous caregiver education and consultation, and with the beginning of intensive speech and language therapy, language and communication skills made progress in a controlled and familiar manner. The special educational treatment made progress in visuospatial coordination and fine motor skills. General psychomotor function showed improvement upon the introduction of target and intensive habilitation treatment, suggesting that there is a connection between habilitation treatments and positive trend in children’s development.

The existing literature confirms the positive impact of specialized habilitation intervention on target skills in children with FAS. With the adapted intervention procedures, according to Peadon et al.₁³, there is a more significant progress in the development of mathematical skills and mathematical memory than in the progress in phonological awareness and fine motor skills. In addition, there is progress in the development of communication skills, verbal comprehension and attention₁³.

Deficits in sensory processing are associated with behavioral disorders in older age. Therefore, the treatment and habilitation intervention should be sensory-based and considerate of this problem₁⁴. It is extremely important to continuously follow up the child’s development to prevent learning ⁵ and behavioral difficulties₁⁰,₁⁶. Based on parental assessment, socio-emotional disorders, attention deficits and aggressive behavior are significant problems in children with FAS. In a recent study, the relationship between social skills and verbal IQ score was assessed in children with FAS and controls. The results suggest that social deficits in children with FAS are beyond what can be explained by low IQ score, indicating that they may be arrested but not simply delayed⁹. In addition, it is indicated that the adaptive functioning of children with FAS is well below expectations when compared with the level of cognitive functioning. However, IQ scores may not reflect the full range of difficulties or extent of functional compromise₁⁷.

The relationship between cognitive and behavioral deficits caused by prenatal alcohol exposure and the structural, ‘brain-based’ changes is determined by use of comprehensive neuroimaging studies. These physical findings confirm the importance of detecting and diagnosing medical condition and recognizing developmental disorders in FAS in order to provide an appropriate medical treatment and habilitation₁⁷.

Early diagnosis of children with FAS is the first step in the early intervention treatments that stimulate skills essential for independence₁⁶. Finding the localization of changes in the CNS allows better intervention strategies⁹. Early diagnosis and a stable, nurturing home environment have been identified as a strong protective factor in persons with FAS ⁴,₁⁸. There is no detailed description of the habilitation strategies in the literature, and the intervention protocols
are not firmly established. Although previous studies have shown significant methodological challenges in the habilitation procedures, there are new programs of early intervention and habilitation appearing in the field19. The intervention procedures should rely on stabilization of the home/family environment and work to improve the parent/caregiver-child relationship4,20.

The involvement of mothers of toddlers with FAS in the early intervention and educational programs can protect children from negligence and improve the mother-child relationship. According to the studies from Canada and USA, the diagnosed children of mothers who participated in the educational and habilitation programs achieved results similar to the children without deficits in cognitive, motor and social development. The mothers also showed considerable progress in staying sober and getting control of their and the child’s life21.

Sensory integration therapy, child’s massage and occupational therapy, and also sucking and feeding therapy in infancy showed positive effect on the development of children with FAS. Raising awareness and educating parents about the child’s needs will bring empathy and stabilization to the parent-child relationship21.

Children with FAS often need specially focused education and healthcare methods because of difficulties in understanding the causes and consequences, as well as in understanding other executive skills that are typical for FAS. Parental education should focus on understanding the difference between the children with FAS and children without developmental deficits, and also it can make them aware of what the child with FAS can achieve with proper habilitation. This can be used as a preventive method for the behavioral and other problems that can occur in adulthood4,18,20.

References

Valentina Matijević et al.

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Sažetak

HABILITACIJSKI TRETMAN DJETETA S FETALnim ALKOHOLnim SINDROMOM – PRIKAZ SLUČAJA

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Fetalni alkoholni sindrom obilježen je trijadom simptoma kao što su facijalne malformacije, prenatalni i postnatalni usporeni razvoj i disfunkcija središnjega živčanog sustava. Posljedica je teratogenog djelovanja alkohola za vrijeme gestacija. Učestalost fetalnog alkoholnog sindroma iznosi 1-3 slučaja na 1000 novorođenčadi. S neurološkog stajališta može doći do oštećenja središnjega živčanog sustava. Strukturna oštećenja uzrokuju odstupanja u razvoju grube i fine motorike, okulomotorike i poteškoće u senzomotornoj integraciji. S funkcionalnog stajališta može doći do složenih spoznajnih poremećaja i poremećaja u ponašanju, poremećaja pozornosti i kontrole impulsa, poteškoća u učenju, socijalnoj percepciji i komunikaciji. Složena simptomatologija ukazuje na disproporcionalni razvoj vještina te zahtijeva timsku dijagnostiku i individualizirano planiranje habilitacijskog djelovanja. U radu se prikazuje studija slučaja dječaka u dobi od 4 godine s dijagnozom fetalnog alkoholnog sindroma. Dječaka je pratio interdisciplinarni stručni tim sastavljen od fizijatra i neuropedijatra, uz polivalentne habilitacijske postupke logopeda, rehabilitatora i psihologa.

Ključne riječi: Fetalni poremećaji alkoholnog spektra; Rehabilitacija; Njegovatelji – izobrazba; Profesionalac-obitelj, odnosi