CASE STUDY

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A PICTURE OF SPEECH DISTURBANCES IN CHILDREN WITH CEREBELLAR ATAXIA

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SUMMARY

Background: The paper presents the logopedic problems of diagnosing children with cerebellar ataxia. Cerebellar lesions, which result inter alia in a discoordination of the muscular movements of the articulatory apparatus, are the cause of ataxic dysarthria – the type of dysarthria least described in the Polish subject literature. The etiology of acquired ataxic dysarthria points inter alia to cerebellar tumors, and neurodegenerative and metabolic diseases, while congenital dysarthria is attributed, for example, to cerebral palsy.

Material/ Methods: The paper presents the results of the speech-therapy examinations of three children aged 6 to 12 years with a medical diagnosis of ataxia during medical conditions causing cerebellar dysfunctions such as cerebral palsy (A), brain tumor (B), and metabolic disorders (C). The descriptive part of the diagnostic procedure takes into account the examinations using *The Dysarthria Scale. Children's Version* devised by U. Mirecka and K. Gustaw, experimental phonological hearing tests, observations of linguistic behaviors, the assessment of the body posture in communication situations, and the assessment of the anatomical condition of the speech apparatus.

Results: The following were diagnosed in the three children: Case A – moderate ataxic dysarthria, Case B – moderate ataxic dysarthria, oligophasia and dysglossia, and Case C – mild ataxic dysarthria and multiple, complex dyslalia. The paper presents the symptoms of ataxic dysarthria at the segmental and suprasegmental levels of the examined children's utterances, both those regarded as typical and less typical.

Conclusions: The conclusions emphasize that speech-therapy practice should take into account, when diagnosing children with cerebellar ataxia, the possibility of other speech pathology entities co-occurring with dysarthria and the occurrence of less typical dysarthric symptoms in children as compared with adults.

Key words: ataxic dysarthria, cerebellar dysarthria, cerebral palsy

BACKGROUND

Ataxia is defined as an "inability to generate a normal or expected voluntary movement trajectory that cannot be attributed to weakness or involuntary muscle activity about the affected joints" (Singer et al., 2010, p. 16). It can appear as a result of lesions of the cerebellum or spinocerebellar tracts; consequently, deficits of cerebellar proprioceptive input and defects of cerebellar processing occur (MacDonald, 1994). Ataxia is also characterized as a disorder of motor coordination and balance; there can be observed discoordination of the muscles of articulation, head, neck, trunk and limb (Lehmann-Horn & Ludolph, 2004). Three basic ataxic syndromes can be distinguished: sensory ataxia, the midline cerebellar syndrome, and the lateral cerebellar syndrome (MacDonald, 1994). ataxia can manifest itself in the lesions of the cerebellum as inaccurate movement to a target (dysmetria), impaired rhythmicity of rapid alternating movements (dysdiadochokinesis), decomposition of multijoint movements (dyssynergia), nystagmus (Prusiński, 1983; Singer et al., 2010). Ataxia can be hereditary or acquired (Sidtis et al., 2011). Ataxia in childhood may be a symptom of various neurologic diseases – dysfunction of the cerebellum occur in acute, intermittent, and progressive disorders (Vedolin et al., 2013). According to the research of Jayaprakash Gosalakkal (2001), the disorders in children ataxia differ from those in adults.

Speech deficits that are caused by the lesions of the cerebellum and its connections are called ataxic dysarthria/cerebellar dysarthria (Folker et al., 2011; Murdoch & Theodoros, 1998). Dysarthria is defined as performance disorders of the motor speech mechanism; neurological impairments – the cause of dysarthria – are manifested in dysfunctions in the respiratory, phonatory and articulatory apparatus that result in distortions of the phonic substance of utterances at the segmental and suprasegmental levels. Dysarthric disorders have a different range and intensity (in extreme cases they consist in the inability to produce speech sounds) and a large group of patients have to cope with a serious problem, which is the limited intelligibility of their pronunciation that makes it difficult or impossible to impart information through the articulatory-auditory channel (Mirecka, 2008, 2012).

The features characteristic of ataxic dysarthria are: prosodic excess (main symptoms are prolonged phonemes, excess and equal stress, prolonged intervals and slowing of verbal expression), phonatory-prosodic insufficiency (in which occur harshness, monopitch and monoloudness) and articulatory inaccuracy (which is associated with the imprecision of consonant production, irregular articulatory breakdowns and distorted vowels) (Brown et al., 1970). Duffy (1995) emphasized a breakdown in the timing and coordination of speech as the most important features in ataxic dysarthria. "Ataxic movements are halting, imprecise, jerky, poorly coordinated and lacking in speed and fluidity or smoothness […] Incoordination and reduced muscle tone appear responsible for the slowness of movement and inaccuracy in the force, range, timing and direction of speech movements" (Duffy, 1995, p. 165, 183).

As Murdoch and Hudson-Tennent (1994) claim, childhood dysarthria (classified as being either acquired or congenital/developmental) is complicated by the interaction between the acquired and developmental components of the disorder. Clinical features which are characteristic for acquired ataxic dysarthria in childhood: muscles – incoordination, movements are slow, inaccurate and irregular; speech – breakdown in articulatory and prosodic aspects of speech. The aetiology of childhood acquired ataxic dysarthria includes posterior fossa tumours, infections, degenerative disorders, toxic, metabolic and endocrine disorders and severe CHI (Murdoch & Horton, 1998). Congenital ataxic dysarthria in childhood occurs for example in cerebral palsy, however, ataxic dysarthria is the least common form of dysarthria in CP.

CASE STUDIES

The paper presents the results of the speech-therapy examination of three children with a medical diagnosis of ataxia during medical conditions that cause dysfunctions of the cerebellum. The following children were selected for the needs of the present article; children who were subjected to medical diagnostic procedures at the University Speech-Therapy Laboratory, Department of Logopedics and Applied Linguistics, Maria Curie-Skłodowska University in Lublin.

Case A

A twelve-year-old girl medically diagnosed with *cerebral palsy – cerebellar form*. Balance disorders were shown as the main symptom of ataxia. Psychological diagnosis: below-average intelligence.

Case B

A ten-year-old girl with a medical diagnosis of *D43.1 Neoplasm of uncertain or unknown character: the brain, subtentorial structures*; coexistent disorders were diagnosed as *G40.8 Epilepsy and epileptic syndromes of an unspecified character* and *G98 Other disorders of the nervous system*. Neuroimaging examinations showed generalized atrophic changes of the cerebellum. Cerebellar symptoms manifesting as ataxia were pointed out. Psychological diagnosis: moderate mental retardation.

Case C

A six-year-old boy with a medical diagnosis at the age of five of *G96.8 Acute cerebellar ataxia (*manifesting in walking disorders, lowered muscular tone, discrete nystagmus) and with a post-diagnostic diagnosis of *E88.9 Other metabolic disorders – Metabolic disorders, undefined.* Normal intellectual development.

The descriptive part of the speech-therapy diagnostic procedure took into consideration the examinations using the *Dysarthria Scale* and experimental tests for phonological hearing (Mirecka, 2013), observation of linguistic behaviours,

assessment of the body posture in communication situations and the anatomical speech apparatus.

In the individual clinical tests of the presented children, the main diagnostic instrument used was the *Dysarthria Scale, Children's Version* by U. Mirecka and K. Gustaw (2006), which is an assessment profile based on an observation of the manner of a patient's performance of individual tasks that involve the speech apparatus; in the analysis of the subject's utterances they are founded on perceptual assessment. It is largely based on S. J. Robertson's *Dysarthria Profile* (1987) and on the *Perceptual speech dimensions* and *Perceptual vocal abnormalities* developed by H. J. Chenery, B. E. Murdoch et al. (Murdoch, 1998)**,** being a modification of these techniques complemented with my own contributions. The *Dysarthria Scale* consists of 70 tasks in nine consecutive spheres, whose arrangement is based on transition from more complex acts and functions to simpler ones: I. Self-Assessment, II. Intelligibility, III. Articulation, IV. Resonance, V. Prosody, VI. Phonation, VII. Respiration, VIII. Alternating movements, IX. Functional condition of the musculature of the articulatory apparatus. Tasks are graded on the 5-point scale (0 to 4): 0 – absence of disorders, 1 – slight degree of disorder, 2 – moderate degree of disorder, 3 – significant degree of disorder, 4 – profound degree of disorder.

RESULTS

The results of examinations carried out using *The Dysarthria Scale. Children's Version* are presented in tables in accordance with the spheres distinguished in *The Dysarthria Scale*, and with the scores obtained by each child (A, B, C).

The twelve-year-old girl with below-average intelligence (Case A) noticed the problem of decreased comprehensibility of her own verbal messages, she also pointed to her breathing difficulties; in contrast, she did not feel fatigability while speaking or phonation problems.

The assessment of the problems connected with her own speech, using the criteria defined in the *Dysarthria Scale*, proved too difficult for the ten-year-old girl with mental retardation (Case B) and the six-year-old boy (case C). The inability to perform these tasks was due to the failure to understand instructions and to difficulties with making self-assessment, which, in turn, was dependent on the level of the mental (mainly conceptual) development of the subjects. Similar problems with self-assessment were found in mentally disabled children and in younger children (aged 6-7) in studies concerning dysarthria in cerebral palsy (Mirecka, 2013).

Table 1. Results of the assessment: sphere I. Self-Assessment

TASK	GRADE		
	A	B	C
1.Assessment of the intelligibility of a patient's own utterances	2	-	-
2. Fatigability during speech	0	-	-
3. Respiration problems	1	-	-
4. Vocal difficulties	0	-	-

In the examiner's assessment, the lowered intelligibility of patients free utterances was at the level of moderate difficulties (ranging from 20 to 50% of unintelligibility); the same intensity of problems was reported in word and sentence repetition tests in the examination of Girl B and Boy C. In contrast, in the case of Girl A, repetition tests came out better in respect of intelligibility, which can be linked to the child's greater self-control in tasks of this type.

In comparison to the other children, better results were scored in the articulation assessment by Girl A: her articulation difficulties were described as moderate, manifesting as small, sporadic vowel deformations, consonant substitutions and deformations (dentalized sounds and /r/), consonant cluster simplifications and as-

Table 2. Results of the assessment: sphere II. Intelligibility

TASK	GRADE		
	A	B	C
1. Intelligibility of one-word utterances while the patient repeats words	1	2	2
2. Intelligibility of one-sentence utterances while the patient repeats sentences	1	2	2
3. Intelligibility of the patient's free utterances	2	2	2

Table 3. Results of the assessment: sphere III. Articulation

TASK	GRADE		
	A	B	C
1. Vowels in words	1	2	1
2. Consonants in words	2	3	3
3. Consonant clusters in words	1	1	2
4. Polysyllabic words	1	2	3
5. Sentences	2	3	3
6. Articulation in free utterances	2	3	3

Table 4. Results of the assessment: sphere IV. Resonance

TASK	GRADE		
	A	B	C
1. Resonance realization in words	1	0	0
2. Resonance realization in sentences	1	0	0
3. Resonance realization in free utterances	1	0	0

Table 5. Results of the assessment: sphere V. Prosody

TASK	GRADE		
	A	B	C
1. Imitation of intonation	1	2	2
2. Intonation in free utterances	1	2	1
3. Imitation of different stress patterns	3	2	2
4. Maintain appropriate rhythm in sentences	1	3	2
5. Maintain appropriate rhythm in free utterances	1	3	2
6. Maintain appropriate rate of speech in sentences	1	1	1
7. Maintain appropriate rate of speech in free utterances	1	1	1
8. Ability to accelerate rate of speech	3	3	2
9. Ability to slow down rate of speech	2	3	2
10. Length of phrases in sentences	2	1	0
11. Length of phrases in free utterances	2	1	0
12. Synchronization of respiration, phonation and articulation in words	1	1	0
13. Synchronization of respiration, phonation and articulation in sentences	2	1	0
14. Synchronization of respiration, phonation and articulation in spontaneous utterances	2	1	0

similations within consonant clusters. In Girl B and Boy C, the intensity of articulation disorders was considerable; in Case B there were deformations and substitutions of both consonantal and vowel phonemes (greater intensity of disorders being reported in consonants), contact (adjacent) assimilations and distant ones, and word structure reductions (including consonant cluster reductions); in Case C, phoneme deformations were not reported; however, there was a considerable intensity of substitutions, particularly in consonants (this covered 17 phonemes) and the accumulation of syntagmatic changes (mainly assimilations and word structure reductions). It should be emphasized that phonemic hearing examined using experimental tests for auditory paronyms differentiation (see Mirecka, 2013) was assessed as normal in each of the three presented cases: the found difficulties cannot therefore be linked to this aspect of auditory processing. However, the articulation of Girl B might have been affected to some extent by an occlusion defect (distoclusion), while in Boy C – by the ongoing exchange of milk teeth for permanent teeth and disorders of auditory-kinesthetic-motor integration.

Abnormal nasal resonance in the form of slight, unstable hypernasality was observed only in Girl A.

Out of the prosodic phenomena examined here, the majority were realized incorrectly, with the intensity of difficulties ranging from slight to considerable. In general, the fewest irregularities in the prosodic organization of utterances were observed in Boy C – there were moderate difficulties with imitating intonation and stress patterns in sentences, with maintaining appropriate rhythm in the repeated sentences and in free utterances (speaking with sound prolongations, chanting and staccato), and with the intentional acceleration and slowing down of the rate of speech, while slight difficulties were reported in the realization of intonation in free utterances (monotonous intonation)and with maintaining the

Table 6. Results of the assessment: sphere VI. Phonation

TASK	GRADE		
	A	B	C
1. Vocal attack – /a/	1	0	0
2. Maximum phonation time of /a/	2	1	0
3. Voice volume during speech	2	1	0
4. Raise voice volume /a/	2	1	0
5. Lower voice volume /a/	1	1	0
6. Pitch of voice	0	0	0
7. Raise pitch /a/	1	2	0
8. Lower pitch /a/	1	2	0
9. Quality of voice	2	1	0

Table 7. Results of the assessment: sphere VII. Respiration

TASK	GRADE		
	A	B	C
1. Respiration at rest	1	1	0
2. Respiration during speech	2	1	0
3. Length of exhalation during emission of /s/	2	1	0
4. Length of exhalation during emission of a series of /s/	2	1	0

appropriate rate of speech (a somewhat slowed down rate); the length of phrases realized on one exhalation and respiratory-phonatory-articulatory synchronization were normal. Girl A had considerable difficulties with imitating stress patterns (despite the fact that in the experimental test of phonological prosodic hearing she correctly discriminated between sentences with different logical stress, and between sentences with different intonation) and difficulties with intentionally accelerating the speech rate; she had moderate problems with intentionally slowing down the rate of speech, with length of phrases, and with respiratory-phonatory-articulatory synchronization (speech on residual air and on inhalation occurred), and she had slight problems with intonation realization (monotonous intonation), with the maintenance of rhythm (speaking with prolongation of sounds, and a slight tendency to chant) and with the speech rate (slowed down). Girl B showed substantial difficulties with intentionally changing the speech rate – both accelerating and slowing it down, and with maintaining an appropriate speech rhythm (chanting, staccato, speaking with prolongation and inappropriate stress); she had moderate difficulties with imitating intonation and stress patterns and

Table 8. Results of the assessment: sphere VIII. Alternating movements (diadochokinesis)

TASK	GRADE		
	A	B	C
1. Open and close mouth rapidly within full range of mandibular movement	2	1	1
2. Purse and stretch lips rapidly	2	3	0
3. Protrude and retract tongue rapidly	2	2	1
4. Raise and lower tongue rapidly outside of the oral cavity	2	3	0
5. Move tongue rapidly to the right and left lip corner	3	4	0
6. Repeat rapidly /u – i/	1	2	0
7. Repeat rapidly /a – y/	2	2	0
8. Repeat rapidly /pa – ta – ka/	1	2	0

Table 9. Results of the assessment: sphere IX. Functional condition of musculature of the articulatory apparatus

TASK	GRADE		
	A	B	C
1. Purse lips	0	3	0
2. Stretch lips	0	2	0
3. Tone of lips	1	1	0
4. Tongue protrusion	1	0	1
5. Tongue retraction	1	1	0
6. Tongue appearance	1	1	0
7. Tongue tip into right cheek	4	4	0
8. Tongue tip into left cheek	4	4	0
9. Move tongue tip to right lip corner	4	4	0
10. Move tongue tip to left lip corner	4	4	0
11. Raise tongue tip inside the oral cavity – to the upper gums	0	4	0
12. Raise tongue tip outside of the oral cavity – to the upper lip	2	4	1
13. Tone of tongue	1	1	0
14. Elevation of soft palate during emission of /a/	0	0	0
15. Elevation of soft palate during emission of a series of /a/	1	0	0
16. Swallowing saliva at rest	0	1	0
17. Swallowing saliva during speech	0	1	0
18. Involuntary movements	0	0	0
19. Facial symmetry at rest	0	0	0

with intonation realization in free utterances, and she showed slight difficulties in the speech rate (slowed down), phrase length and with respiratory-phonatory-articulatory synchronization (speaking on residual air). It should be stressed that Girl B was not able to understand the instructions in the phonological prosodic hearing test, which is why this area was not assessed.

In phonatory tests, Girl A scored the worst: there were moderate problems with phonation prolongation, with the voice volume (low voice), with intentionally increasing the voice volume, and also with its quality (weak and hoarse); slight dysfunctions were observed in the vocal attack (a tendency for breathy attack), in intentionally raising/lowering the voice pitch and in lowering the voice volume; the voice pitch being normal. Girl B had moderate difficulties with intentionally raising/lowering the voice pitch, and slight difficulties with phonation prolongation, loudness of utterances (unstable voice volume) and with the voice quality (hoarse); the vocal attack and voice pitch were normal. No abnormalities in using the voice were found in Boy C.

Respiratory tests showed the regular functioning of the respiratory apparatus in Boy C, slight respiratory dysfunctions in Girl B (shallow respiration at rest, irregular respiration with a short expiratory phase during speech, and somewhat reduced attempts to intentionally prolong expiration), and moderate disorders of respiratory functions in Girl A (the same type as in Case B but with a greater intensity).

In alternating movements tests no problems with diadochokinesis, apart from slight difficulties with the performance of tasks 1 and 3, were found in Boy C. The two girls (A and B) showed disorders in this sphere; both in motor tests of the speech organs (tasks 1-5) and in the tests for pronunciation of sounds and syllables contrasted in articulatory terms (tasks 6-8). A greater intensity of problems occurred in Girl B (inter alia she was unable to move the tongue from one lip corner to the other). In both Cases A and B the movements were slowed down, inharmonious, and not carefully executed.

The assessment of the functional state of the muscles in the articulatory apparatus showed, in the case of Boy C, only slight difficulties in two tasks: tongue protrusion (asymmetrical arrangement) and its elevation onto the upper lip. In contrast, Girls A and B had problems with most tasks that assessed the performance of the articulatory apparatus, the intensity of the problems being highly varied. Both of them were unable to move the tongue towards the lip corners or push out the cheeks with the tongue; furthermore, girl A was not able to raise the tongue upwards or push out the cheeks with the tongue, while Girl B was unable to raise the tongue upwards (to the upper gums and the upper lips). Girl A correctly raised the tongue to the upper gums but she had moderate difficulties with raising the tongue onto the upper lip. Generally speaking, more difficulties with moving the lips and the tongue were reported in Girl B, she also had an infantile type of swallowing. It should be noted that muscle tone disorders within the lips and the tongue were observed in Girl A (the unstable tone) and in Girl B (the decreased tone).

When assessing non-verbal communication behaviours, no abnormalities in the body posture and in visual contact were observed.

CONCLUSIONS

The analysis of the collected data and their interpretation, taking into account specialist examinations (medical, psychological and pedagogical, derived from the documents made available) were the basis for the logopedic diagnosis.

In order to determine the degree of intensity of dysarthric disorders, the following distinction was referred to: *mild, moderate, severe, and profound dysarthria*; in this division the term *profound dysarthria* is applied to the case of the maximum intensity of dysarthric symptoms leading to extremely unintelligible pronunciation or the impossibility of producing speech sounds (Mirecka, 2013).

Case A – a twelve-year-old girl with the cerebellar form of infantile cerebral palsy: moderate ataxic dysarthria was diagnosed.

Case B – a ten-year-old girl with generalized atrophic changes of the cerebellum: moderate ataxic dysarthria, oligophasia (the low level of linguistic and communicative competence and skills) and dysglossia (determined by an occlusion defect) were diagnosed; dysphagia being shown as a co-occurring disorder.

Case C – a six-year-old boy with an earlier diagnosis of acute cerebellar ataxia and subsequently a diagnosis of undefined metabolic disorders: mild ataxic dysarthria and multiple, complex dyslalia (determined by disorders in auditory-kinesthetic-motor integration) were diagnosed.

The picture of speech disorders presented is made up of dysarthric symptoms at the segmental and suprasegmental level of utterances (in Cases A, B and C), associated with lesions/changes within the cerebellum and cerebellar tracts, overlapped by the symptoms of co-occurring speech disorders: oligophasia and dysglossia (Case B) and dyslalia (Case C).

The most important symptoms of ataxic dysarthria in the children at the segmental level of utterances are paradigmatic disorders: deformations and substitutions of consonantal phonemes as well as syntagmatic disorders: word structure reductions (mainly simplifications of consonant clusters) and sound assimilations. The dominant symptoms at the suprasegmental level were speech rhythm disorders (speech with prolonged sounds, a tendency to chant and staccato), speech rate disorders (the slow speech rate associated with slower articulation of sounds and sometimes with their prolongation), intonation disorders (monotonous intonation), and – in the examined girls – phonatory insufficiency (hoarseness, wrong voice volume). What should be regarded as less typical symptoms of ataxic dysarthria observed in the studied girls were the observed disorders of respiratory-phonatory-articulatory synchronization (speech on residual air, and also speech on inhalation reported in one of them), shortened phrases, and hypernasality (in one of the subjects).

The symptomatology of dysarthric disorders in children, as shown by researchers (e.g. Obrębowski & Woźnica, 1997; Love, 2000), is less typical and less pronounced than in adults (see e.g. Jauer-Niworowska et al., 2014). In the studies by van Mourik et al. (1998) in a group of children after the resection of the cerebellar tumor, acquired ataxic dysarthria manifested not only in inaccurate

articulation and speech slowdown (which was the main symptom) but also in hypernasality in some children: these authors contend that Darley's clinical classification of dysarthria, with a list of symptoms typical of the dysarthria types distinguished in it, is a good instrument for describing dysarthria in adults, but it does not turn out to be useful inthe differential diagnosis of dysarthric disorders in children. The results of our own studies given in the present article and the results of studies conducted in a group of 36 dysarthric children with infantile cerebral palsy (Mirecka 2013) confirm the observations of the cited authors.

The most general conclusions for speech-therapy practice that can be drawn from the presented investigations are as follows:

In diagnosing children with cerebellar ataxia it is necessary to take into account the possibility of the co-occurrence with dysarthria of other speech pathology entities, which may hinder an analysis of disorder manifestations at the phonic level of patients' utterances, and which requires accurate examination of the immediate causes of the observed difficulties.

It is necessary to take into account the possibility that less typical symptoms will occur in children when compared with adults.

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