Acquired Neurological Mutism and Acquired Dysarthria in Childhood

Acquired Neurological Mutism and Acquired Dysarthria in Childhood

Verworven neurologisch mutisme en verworven dysarthrie bij kinderen

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De gedachte vliegt en de woorden
gaan te voet. Ziedaar het hele
drama van de schrijver.

(J. Green, 1974)

Aan mijn ouders en mijn kinderen

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List of abbreviations

ACD	Acquired Childhood Dysarthria
ANCM	Acquired Neurological Childhood Mutism
CT	Computer Tomography
GCS	Glasgow Coma Scale
MRI	Magnetic Resonance Imaging
MSD	Mutism and Subsequent Dysarthria
VP	Ventriculoperitoneal

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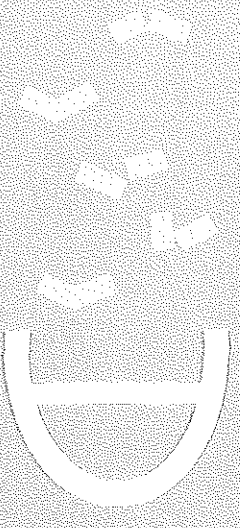
General introduction and study outline



1.1. Introduction

This thesis comprises neuropsychological studies on a syndrome which may occur in children after a neurosurgical operation, in which a mass lesion, located in the cerebellum is resected. In the immediate postoperative course the children awake from anesthesia and start to speak. During the next few hours or days a complete loss of speech – mutism – occurs. This dramatic postoperative complication is not necessarily associated with other new neurological deficits. The mute phase may vary in duration and is followed by a period characterized bij a severe motor speech problem – dysarthria –.

The first description was given by Rekate et al. [1] a decade ago. The dysarthria itself did not baffle the authors – as this had been known for a long time [2] – as much as the mutism: the authors reported that they had not been able to locate a description of this most severe derangement of speech after cerebellar tumor resection [1]. Since then, the syndrome of 'mutism and subsequent dysarthria' after cerebellar tumor resection (MSD) has been described in a number of case studies and review studies. In 1990, 18 cases were reported [3], and until now approximately 70 cases with MSD have been published all over the world [see reviews 4-6]. It has been estimated that it occurs in 15-20% of all cases with cerebellar tumor surgery.

The characteristics of this 'new postoperative clinical syndrome' [7] are: it occurs almost exclusively in children; the mutism may last for days to months, but is of a transient nature; the dysarthria disappears gradually and (almost) completely.

All studies have attempted to find explanations for the occurrence of MSD. There seemed to be a relation with the tumor type, as medulloblastoma was more frequently associated with MSD than any other tumor type [4,6,8]. The surgical procedures were considered in connection with the occurrence of MSD, investigating the possible influence of damage to the dentate nuclei, of intense manipulation of the brainstem during surgery, and of air embolism during operation causing ischemic lesions and edematous lesions [6,9-13]. The occurrence of postoperative hydrocephalus and infection were also considered of importance [3,8]. The anatomical and neurophysiological substrate for MSD was considered to be either exclusively cerebellar [1,9,12,14,15] – dentate nuclei, vermis and cerebellar hemispheres – or more diffuse [3,4,6,13] – brainstem and/or cerebellar peduncles –, or the dysfunction of dentato-thalamo-cortical fiber bundles [8,11,16]. Non-organic explanations were also considered [3,13,14]: negativism on the part of the child, who felt betrayed after awakening in the intensive care unit, resulting in anxiety, whining, food intake refusal, and in a quick recovery of speech as soon as the child was at home [3]. These far from negligible psychic factors were considered of importance and it was thought that they could retard the onset of speech.

Thus, when we started our studies there were many pathophysiological possibilities but no conclusive explanations for MSD. Moreover, there were no neuropsychological studies describing the underlying mechanism of the mutism and the subsequent

dysarthria. The mutism was generally considered the most severe form of dysarthria. Other studies [3,9,17] suggested an 'apraxia', a loss of the ability to carry out those movements required for speech. However, neurobehavioral features that characterized the evolution of the mute state and which could predict the onset of speech were unknown. The dysarthria was frequently labeled cerebellar, referring to speech features in adults with cerebellar damage, but speech features had not been studied in detail.

1.2. Outline of the study

Our studies were carried out at the Sophia Children's Hospital Rotterdam. From 1989 to 1996, 42 children underwent a cerebellar tumor resection, 12 of whom suffered from MSD postoperatively. We had the opportunity to examine these children with the following aims in mind:
1) to describe the neuropsychological aspects of mutism and to detect factors which could predict the onset of speech;
2) to analyze speech features of the subsequent dysarthria.

Alle children were examined prospectively according to a standard protocol. Examinations were audio- and videotaped in order to make reliable judgments of the behavioral features, the orofacial movements and the speech features.
Of the 12 children, we excluded two three-year-old patients from further analysis. There was no doubt that they suffered from MSD. However, reliable evaluation of postoperative dysarthric features was not possible, as preoperative speech was scarce and revealed articulation problems characteristic of immature developing speech.

Our studies focus on different aspects of MSD. In **Chapter 2**, which was published in 1994, and in **Chapter 3**, which contains an update until 1997, we present data on 10 children who suffered from MSD. Risk factors for the occurrence of MSD are pointed out. Neuropsychological observations in the MSD children served as a starting point for further studies on the mutism and the dysarthria: the observations suggested that during the mute phase MSD children appeared to have difficulties carrying out voluntary complex orofacial movements. As to the dysarthria we were at that time unable to identify a distinctive cluster of speech features.

In **Chapter 4** we compare the new form of mutism in MSD with other well-recognized types of mutism. We have analyzed behavioral features in one MSD case and in three other children, who became mute after damage in other brain structures due to various etiologies. Basing ourselves on a systematic analysis of behavioral aspects, summarised in an examination protocol, we show that mutism in childhood has many 'faces', and types of mutism differ from each other in neurobehavioral aspects. MSD differs from other types of mutism such as the akinetic state, from aphasic mutism and from mutism due to auditory agnosia. Impairment of voluntary orofacial movements seemed to be the most significant disturbance in MSD.

We then carefully analyzed mutism in MSD. For hospital staff and even for parents the disappearance of the mutism and the onset of speech always came unexpectedly. Factors which could predict the recurrence of speech were poorly understood. Therefore, we closely followed five children throughout the mute phase and shortly after the onset of dysarthria and we analyzed the role of simple and complex orofacial movements. These data are presented in **Chapter 5**.

Before analyzing the dysarthric speech features of MSD, we made a survey of the literature on speech features of Acquired Childhood Dysarthria, henceforward referred to as ACD. **Chapter 6** presents a review of studies on ACD. It shows that dysarthria following posterior tumor resection has received more attention than any other type of dysarthria. Most pediatric studies refer to the adult classification of dysarthria [18], thus implicitly assuming the validity of this adult model. This assumption is hazardous, as normal speech development may reveal speech features which are considered pathological in adults [19]. The survey shows that dysarthria following cerebellar lesions was studied in approximately 20 children. On the basis of the review one can not conclude whether the speech features in MSD constitute a specific type.

Although most studies on MSD suggested that the dysarthria was cerebellar and resembled the adult cerebellar dysarthria, this was not ascertained by detailed analysis. Moreover, as the necessary conditions for MSD to occur are not exclusively cerebellar [8], this might influence the dysarthria. We therefore carried out an analysis of dysarthria in children with similar etiology (tumoral) but sites of the tumors were different (brainstem vs. cerebellum). These data are presented in **Chapter 7** and show that in spite of overlapping motor impairments, the two groups show different speech features.

1.3. Synopsis of the patient group

Patients were selected on the basis of inclusion criteria stated in the studies presented in Chapters 2, 3, 4, 5, and 7. In Table 1.1 we present a synopsis of the entire patient group and indicate how they are referred to in the chapters.

Table 1.1. Synopsis of the patient group referred to in this thesis.

Patient	Sex/Age	Chapters 2	3	4	5	7
H.	M/6	C1		-	-	A3
G.	F/8	C2		-	3	A2
D.	M/8	C3		-	4	A1
M.	M/5	C4		Case 4	5	A4
Y.	M/4	C5		-	-	-
D.	M/12	-	C6	-	-	A5
N.	M/12	-	C7	-	-	A6
T.	M/7	-	C8	-	-	-
B.	M/17	-		-	1	-
B.*	M/8	-		-	2	-

M = male; F = female; * = mute after recidive tumor resection

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2

The syndrome of 'cerebellar' mutism and subsequent dysarthria

Neurology 1994;44:2040-2046

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C. E. Catsman-Berrevoets
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Abstract

'Cerebellar mutism' refers to a specific childhood disorder in which a complete but transient loss of speech, followed by dysarthria, occurs after removal of a cerebellar tumor. We present a consecutive series of 15 children with this disorder, which we prefer to designate 'mutism and subsequent dysarthria'. The conditions in which it develops suggest also an extracerebellar component of cerebellar mutism. Hydrocephalus at presentation, localization of tumor adjacent to the fourth ventricle, and postsurgical edema of pontine tegmentum are involved in its development.

2.1. Introduction

In children, mutism frequently occurs after severe head injury [1] and in the acute stage of aphasia [2,3]. A number of studies focus on an intriguing clinical picture that occurs after removal of a cerebellar tumor, i.e. mutism and subsequent dysarthria (MSD), commonly labeled 'cerebellar' mutism. As of now, 36 cases are reported in the literature [4-15]. MSD is an iatrogenic complication usually occurring in children after removal of cerebellar medulloblastomas, astrocytomas, and ependymomas. The tumors are large and invade medial cerebellar structures. An associated hydrocephalus, edema, and postsurgical meningitis are regarded as risk factors for the mutism [6,11]. The literature confirms that 90% of the children with MSD are less than 10 years old [4-15]. The youngest patients described were 2 years old [8,12].

The speech characteristics of the subsequent dysarthria are not delineated. The symptom of acquired dysarthria in children receives far less attention than that of acquired aphasia [16-19]. Often the symptom is described qualitatively only as *slurred, slow,* and so forth. Attempts to elucidate cerebellar speech motor control are carried out mainly in adults. Based on a study of 122 patients, most with cerebellar tumors, Lechtenberg and Gilman [20] observed that dysarthria developed most frequently after damage extending into the paravermal segments of the left cerebellar hemisphere and, more specifically, into its superior portion. Amarenco et al. [21] confirmed this in a patient with an isolated cerebellar dysarthria (aprosody with explosive staccato and scanning speech) following a small infarction of the lobulus simplex and lobulus semilunaris superior in the left paravermal zone; they considered a minute mirror-image lesion to be of no importance. Ackermann et al. [22] did not corroborate the notion of an exclusive left-sided paravermal cerebellar speech cortex; three of the four dysarthric subjects in their study had unilateral right-sided ischemia due to occlusion of the superior cerebellar artery. In one patient [22], the lesion was very small and limited to the cerebellar cortex, leading the authors to conclude that the site of the lesion is more important than the size. In none of the speech-impaired patients in Ackermann et al's [22] study could the dysarthria be classified as cerebellar [23]. Darley et al. [23] proposed a cluster of the five most important features of cerebellar dysarthria in the following rank order of frequency: (1) imprecise consonants, (2) excess and equal stress, (3) irregular articulatory breakdown, (4) distorted vowels, and (5) harsh voice quality. Of these features, *excess and equal stress* and *irregular articulatory breakdown* are considered specific for cerebellar dysarthria. In contrast, *imprecise consonants* as a deviant speech dimension is aspecific because it is present in all other types of dysarthria. However, Kluin et al. [24] do not completely agree with Darley's rank order. They consider *excess and equal stress* and *articulatory breakdown* to be specific features as well, but add *alternating loudness, variable rate,* and *fluctuating pitch* as the other frequently occurring features.

We report on a consecutive series of 15 children in whom a posterior fossa tumor was removed. Our aim was (1) to determine the conditions under which MSD develops, (2) to analyze the features of the mutism and the subsequent dysarthria, and (3) to define a more exact localization of the dysfunctional area of the nervous system related to the speech deficit.

2.2. Methods

In three years, 15 children with a diagnosis of posterior fossa tumor were surgically treated in our hospital. All children (10 boys and five girls, ages two-and-a-half to 13 years) had normal psychomotor development. No speech problems immediately before admission had been observed by the parents. In all children, speech was assessed according to the Mayo Clinic Lists of Darley et al. [23]. For rating the severity of the dysarthric components from videotaped samples of spontaneous speech, we used the University of Michigan rating system [24], which extends form 0 (unaffected) to 3 (severely affected). Speech behavior was recorded on videotape before surgery, immediately after surgery, three days after surgery, and, in case of a speech disorder, every second week until recurrence of speech. In those patients, speech behavior was subsequently videotaped at intervals as short as possible for two to four weeks. The follow-up lasted until (nearly) complete recovery of speech (four to 16 months). Neurological examination of the speech musculature was carried out according to the protocol for studying acquired childhood dysarthria [17,19].

2.3. Patients

We divided the patients into three groups (Table 2.1). Group A had no speech problems after surgery and consisted of eight patients. Group B (two patients) did not have mutism but did have mild speech problems after surgery. Group C consisted of five patients with mutism and dysarthria after surgery.

Patient B1. A-10-year-old boy was admitted with headache, vomiting and four days of double vision. Neurological examination was normal except for a position-evoked nystagmus. MRI (Figure 2.1., B1) showed a tumor in the fourth ventricle, which was hypointense on T_1- and hyperintense on T_2-weighted images and enhanced homogeneously with gadolinium. The tumor invaded the right cerebellar hemisphere and the right cerebellar peduncle and extended by way of the foramen of Luschka onto the right prepontine cistern. The tumor (ependymoma grade III) was macroscopically removed completely. After surgery, the boy obeyed simple commands but only started to speak two days after extubation. His voice was very soft, but no other speech abnormalities were heard. At neurological and laryngoscopical examination, a right-sided paresis of the tongue, palatum, and vocal cords were observed. All symptoms disappeared within two weeks.

Patient B2. This four-year-old girl presented with progressive trembling of the hands and insecure gait. At neurological examination, she had a slight intention tremor of the hands and a mild ataxia of the limbs. On CT and MRI (Figure 2.1., B2a and B2b), a very large extra-axial, partly calcified contrast-enhancing tumor was visible, situated ventrally in the posterior fossa. On angiography, the tumor was situated anterior to the basilar artery and was fed form the meningeal truncus of the arteria pharyngea ascendens on the right side. In agreement with a diagnosis of meningioma, an intense tumor blush occurred. An attempt was made to embolize the

solitary feeding artery after hyperselective catheterization, but this failed. At surgery, the greater part of a meningioma was removed.
After surgery, the girl had a flaccid tetraparalysis and respiratory insufficiency for which full ventilation was required. She was alert and communicative. She could protrude her tongue past her teeth and carry out slow alternating movements of the tongue. Although she remained fully dependent on pressure-controlled ventilation, she answered questions by silently moving her lips, and she was able to whisper a limited number of words through a leaking cough. Five months after surgery, her clinical condition remained unchanged. On control MRI, ischemic lesions were present in the ventral part of the lower medulla (Figure 2.1., B2c). Six months after surgery, she died of respiratory complications.

Patient C1. A six-year-old boy presented with diplopia and progressive clumsiness. He had bilateral papilledema, a first-degree nystagmus in all directions, mild bilateral abducens pareses, and a slightly clumsy gait. CT (Figure 2.2., C1) revealed a large mixed hypodense-hyperdense moderately enhancing tumor, partially occupying the fourth ventricle, and an associated hydrocephalus. After insertion of a VP shunt, the neurological signs disappeared. For the first 24 hours after removal of the tumor (a medulloblastoma), the boy answered questions with three- to four-word sentences in a normal voice. He subsequently became increasingly apathetic and did not utter a word. The only sound he produced was a soft and continuous whining. He also did not try to communicate in any nonverbal way except for vigorous stimulation. He had difficulties with swallowing liquids and chewing food. He had a global pyramidal paresis of the right arm and leg and a severe bilateral limb and trunk ataxia (Table 2.1.). Paroxysmal tonic upward deviation of the eyes was noted. During the period of mutism, complex movements of the mouth were severely impaired, but no paresis of facial or pharyngeal musculature or of vocal cords was present (Table 2.2.). He started to recover complex movements of the mouth after eight weeks postsurgery, which slowly improved to almost normal over the next eight months. The ataxia and right-sided paresis gradually disappeared in the course of the eight months. CT six months after surgery showed a wedge-shaped surgical lesion in the midline of the cerebellum.
Recovery of speech. Quite unexpectedly, from five weeks after surgery on, he produced some phonemes, which at seven weeks after surgery were laboriously uttered and unintelligible. Eight weeks after surgery, the child was able to repeat not only words but also short sentences. He spoke with audible respiration {2} (numbers between braces indicate the severity of the dysarthric components according to Kluin et al [24]). His voice was hoarse {2} and nasal {2}, and its volume {2} rapidly decreased. The vowels were abbreviated when pronounced {2}, and consonants and clusters of consonants {3} caused severe articulation disturbance. His articulatory ability gradually improved, as did his voice and breathing, but 26 weeks after surgery, pronunciation of vowels {1} was still too brief. Clusters of consonants were laboriously and slowly uttered {2}. Sometimes a final sound was omitted {1} – for example, the p in the word stop. Expiration was still audible {1}. He was able to maintain sufficient (but harsh) voice volume to complete the speech examination, which demonstrated a slow rate of speech {2}, pauses {1}, very mild nasality {1}, and

Table 2.1. Clinical data of the patients from whom a posterior fossa tumor was removed*

No.	Sex	Age(yr)	Tumor type	Lesion site	Cerebellar signs			Brainstem signs	
					Trunk	Limb	Oculo	Pyramidal tract	Cranial nerves
A. Patients without speech problems after surgery									
A1	F	11	Astrocytoma I	L	(N)N	(+)+	(N)N	(+)N	(VI)N
A2	M	8	Astrocytoma II	L	(N)+	(N)+	(+)N	(N)N	(II)II
A3	M	2	Astrocytoma II	L	(N)++	(+)++	(N)N	(N)N	(VI)VI
A4	F	6	Astrocytoma I	L	(N)+	(+)+	(N)N	(+)N	(N)N
A5	M	5	Astrocytoma I	R	(N)N	(N)+	(N)N	(N)N	(II)II
A6	M	13	Astrocytoma II	R	(N)N	(+)+	(N)N	(N)N	(N)N
A7	F	3	Astrocytoma II	R	(++)++	(++)++	(+)+	(++)++	(II,VI)II,VI
A8	M	5	Ependymoma	L	(N)N	(N)+	(N)+	(N)N	(N)N
B. Patients with mild speech problems after surgery									
B1	M	10	Ependymoma III	R	(N)N	(N)+	(+)N	(N)+	(N)IX,X,XII
B2	F	4	Meningioma	NA	(N)N	(+)NA	(N)N	(N)++	(N)N
C. Patients with mutism and subsequent dysarthria after surgery									
C1	M	6	Medulloblastoma	Medial	(N)++	(+)++	(+)++	(N)++	(VI)VI,XII
C2	F	8	Medulloblastoma	Medial	(N)++	(+)++	(+)+	(N)++	(VI)VII,XII
C3	M	8	Medulloblastoma	Medial	(N)++	(N)++	(N)+	(N)++	(VI)XII
C4	M	5	Medulloblastoma	Medial	(N)++	(+)++	(+)N	(N)++	(N)N
C5	M	4	Ependymoma	R	(N)++	(N)++	(N)++	(N)++	(N)VII,XII

* The results of neurological examination before surgery (parentheses) and immediately after surgery (no parentheses) are presented.

L Left
R Right
NA Not applicable
N No abnormalities
\+ Mildly abnormal
++ Severely abnormal

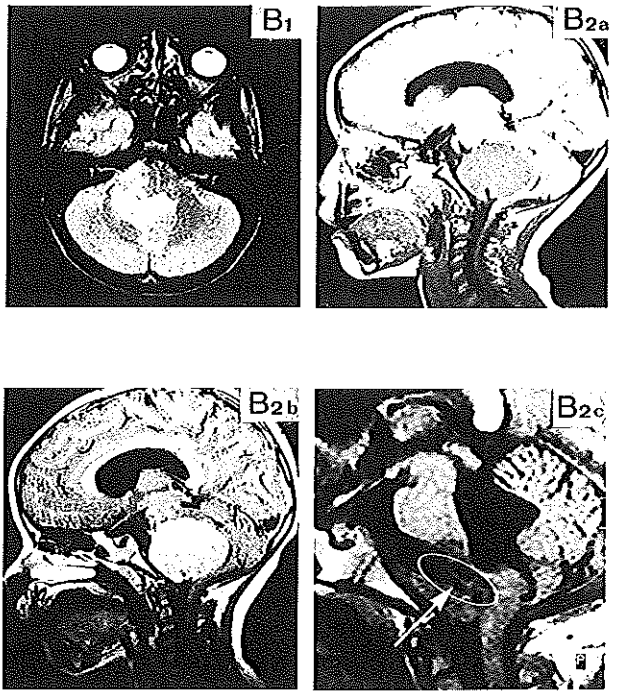


Figure 2.1. MRI of the tumors of group B patients. B1 shows a transverse T2-weighted image with long relaxation time of an ependymoma grade III. B2a and B2b show sagittal T1-weighted images before (B2a) and after (B2b) enhancement with gadolinium of a posterior fossa meningioma in a 4-year-old girl. B2c shows a sagittal T1-weighted image of the same patient after surgery. Note the extensive hypointense area of infarction in the ventral medulla (white arrow).

monopitch [1]. Aphasic features such as paraphasias, neologisms, and syntactic errors were never noted. Language comprehension, as measured by the Test for Reception of Grammar [25], was consistent with age.

Patient C2. An eight-year-old girl was admitted with headache, vomiting, diplopia, and gait disturbance. She had bilateral papilledema, abducens pareses, a paresis of upward gaze, first-degree nystagmus in all directions, and ataxia of the left arm and leg. CT and MRI (Figure 2.2., C2) showed a large contrast-enhancing vermian tumor and hydrocephalus. A VP shunt was inserted and in a second session a medulloblastoma was completely removed macroscopically. After surgery, a slight left hemiparesis was noted. Speech remained unremarkable until two days after surgery, when consciousness deteriorated. A posterior fossa epidural hematoma was diagnosed and evacuated. After removal of the hematoma, a left abducens and left peripheral paralysis of the facial nerve as well as a severe tetraparesis was observed. She produced a soft crying sound but did not talk. She was unable to move her tongue. Swallowing and complex movements of the mouth were impossible despite

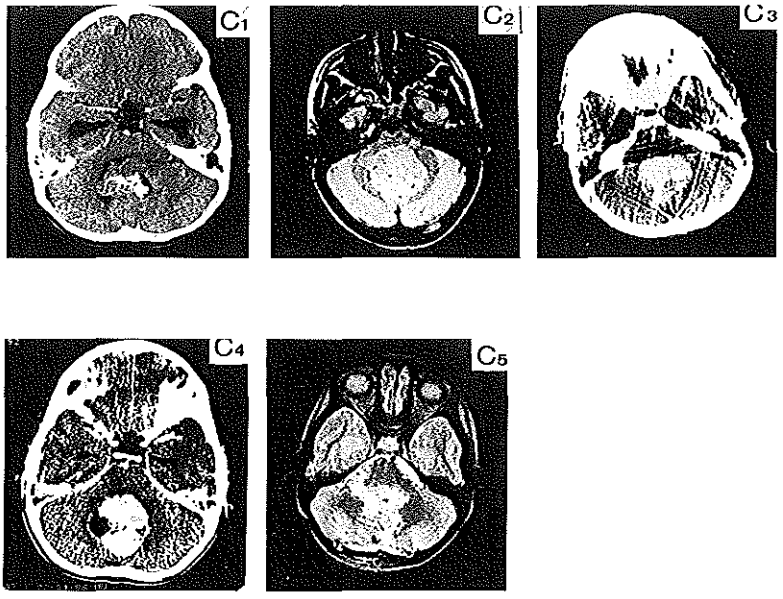


Figure 2.2. Contrast-enhanced CT (C1, C3, and C4) and T2-weighted MRI (C2 and C5) images of the tumors of the group C patients. Those in C1 through C4 are medulloblastomas. C5 shows an ependymoma grade II-III.

an intact seventh cranial nerve on the right side and intact ninth and tenth cranial nerves on both sides. She was severely apathetic but responded to questions adequately by squeezing with the right hand and, later, with the help of a signboard. In the following month the limb pareses and the ataxia slowly subsided.

Recovery of speech. Four weeks after surgery she obeyed simple verbal commands but did not speak. Five weeks after surgery, laughing became audible {2} and sounded like hiccups. She was able to repeat a limited number of phonemes in a soft voice {3} but could not repeat monosyllabic words. After eight weeks, the volume of her voice had increased {2}, she spoke with great effort and with alternating loudness {2}. Her voice was clear. She could shout only very softly {2} and trailed off into a dry cough. When she repeated phonemes and words, the vowels sounded flat {2}. Articulation was poor when repeating simple sentences, which she uttered very quickly. Clusters of consonants were still pronounced with difficulty {2}. Twelve weeks after surgery, her speech rate, as well as the prosody of spontaneous speech, were normal, but she still had mild articulatory difficulties in pronouncing clusters of consonants {1}. Aphasic disturbances were never observed. Further follow-up was not possible because of neurological deterioration due to multiple intracranial metastases of the medulloblastoma in the 20th week. She died six months after surgery.

Patient C3. An eight-year-old boy was admitted with headache after a minor trauma and one day of confusion. He was somnolent, and he had a right abducens paresis

Table 2.2. Results of neurological examination of lower cranial nerves during the initial stage of dysarthria

Symptom (cranial nerve)	Patient C_1	Patient C_2	Patient C_3	Patient C_4	Patient C_5
Trigeminal weakness (V m)	N	N	N	N	N
Sensation (V s)	N	N	N	N	N
Facial weakness (VII R/L)	N/N	N/++	N/N	N/N	+/N
Pharyngeal weakness (IX)	N	N	N	N	N
Lingual weakness (XII)					
Tongue protrusion	+	+	+	+	+
L/R movements	N	+	+	+	+
Complex movements					
Alternating tongue movements	++	++	+	+	++
Clicking	++	+	N	N	++
Chewing	N	N	N	N	N
Swallowing	N	N	N	N	N
Blowing	++	+	N	N	+
Coughing	+	N	N	N	N

m Motor branch
s Sensory branch
R Right
L Left

N No abnormalities
+ Mildly abnormal
++ Severely abnormal

and bilateral papilledema. CT (Figure 2.2., C3) showed a mixed hyperdense-hypodense contrast-enhancing vermian tumor and hydrocephalus. A VP shunt was inserted, and four days later a large medulloblastoma was almost completely removed. A film of tumor remained on the floor of the fourth ventricle. After surgery he spoke words in a normal voice. One day later, he ceased speaking, and a slight paresis of the right arm and severe ataxia of the limbs and trunk was noted (Table 2.1.). CT and spinal fluid examination did not reveal a specific cause for the deterioration. On the fourth day after surgery he developed pneumonia and a respiratory insufficiency for which he required full ventilation for 10 days. Two weeks after surgery, the VP shunt had to be removed because of infection. Three weeks later it was reinserted because of clinical complaints of raised intracranial pressure and widening of the ventricles on CT. One week after removal of the medulloblastoma, he obeyed simple commands. A further week later, after extubation, he was unable to speak but produced a soft crying sound. Chewing and swallowing were unimpaired (Table 2.2.). Four weeks after surgery he did not speak and could be stimulated to cooperate only with great effort. Communication was possible by means of a communication board. On request, the boy wrote the names of objects and answered by yes or no to questions. During the period of the mutism, the function of the cranial nerves was

unimpaired, with the exception of poor tongue protrusion and lateral movements of the tongue. The duration of the mute period was eight weeks. When speech recurred, he had a moderately severe ataxia of trunk and limbs such that he could sit and walk with support.

Recovery of speech. On examination one day after he started to speak again his voice was hoarse {3} and very soft {3}. Respiration was noisy {2}. Only with extreme effort could he produce monosyllabic words, in which the final phoneme was frequently omitted {2}. Ten weeks after removal of the medulloblastoma, his voice was louder and less hoarse {2}. He spoke spontaneously in sentences in a monotonous {1} and nasal {1} way. There were many pauses {2} and forced inspiration and expiration {2}. The articulation of consonant clusters was imprecise {2}. Twelve weeks after surgery there was a strong increase in volume {1}. The pauses had decreased {1}, and inspiration and expiration {1} now matched with slow rate of speech {2}. Difficulties in producing consonant clusters {1} were apparent only when repeating polysyllabic words. At follow-up 16 months after surgery, speech was normal except for a slow rate of speech, which was exclusively heard by his mother when he was tired.

Patient C4. A five-year-old boy was admitted with early morning vomiting, diplopia, and staggering gait. Bilateral papilledema, a vertical skew deviation of the eyes, and a slight ataxia of the left arm and leg were found. On CT and MRI (Figure 2.2., C4), a vermian tumor of mixed density, with some small calcifications and several cystic components filling up the fourth ventricle, and an associated hydrocephalus were observed. After insertion of a VP shunt only papilledema remained. In a second session, a medulloblastoma was macroscopically completely removed. Following surgery, he had a severe ataxia of trunk and limbs, and he was extremely apathetic. He spoke only a few words, after which he did not speak for eight weeks. During this period he obeyed commands willingly, but he cried with a soft whining voice when he was stimulated to answer questions. Cranial nerves were intact except for lingual weakness (Table 2.2.). Complex movements of the mouth and tongue were impaired, but he had no problems with eating or drinking. Two weeks postsurgery, 3 x 5 mg bromocriptine was administered for two weeks, without any effect on recovery of speech.

Recovery of speech. Eight weeks after surgery he started to speak single words. His speech rate {3} was very slow; the words were 'spelled' sound by sound with a strained – strangled voice {2}. Ten weeks after removal of the medulloblastoma, his voice was still strained-strangled {1}; sometimes it was sufficiently loud and on other occasions he whispered {2}. With great effort, he uttered a few words slowly. There was audible inspiration {2} when he repeated two- or three-syllable words. There were prolonged intervals and phonemes {2}. Three months after surgery, spontaneous speech was still severely limited. However, he was able to repeat sentences slowly. Phonemes were still prolonged {1}. After follow-up at four and six months, no abnormalities of speech were noticed except for a slightly slow rate of speech {1}.

Patient C5. A four-year-old Moroccan boy had complaints of headache and early morning vomiting of two months' duration. The parents denied aberrant speech or walking pattern. He was admitted when he suddenly lost consciousness and showed

extension spasms. On MRI (Figure 2.2., C5) a large tumor was seen of mixed density on T_1-and T_2- weighted images, which enhanced in a nonhomogeneous manner with gadolinium. The tumor completely filled the fourth ventricle and extended into the right cisterna magna and dorsally along the medulla and spinal cord down to the level of C-2. Adherence of the tumor was noted at the mesencephalon at the level of the inferior colliculus and at the vermis cerebelli. There was a large proximal hydrocephalus. Consciousness cleared after insertion of a VP shunt. He had bilateral papilledema but no further neurological abnormalities. After removal of the largest part of the tumor (ependymoma grade II-III), he answered questions and according to his parents, he spoke with a normal voice in his Berber dialect. He had a severe paresis of the limbs and a severe ataxia of the right arm. He could not sit unsupported. There was a slight right peripheral facial nerve paresis. He could bring his tongue past his teeth but could not lick his lips. One day after surgery, he became increasingly apathetic and stopped speaking. He developed a respiratory insufficiency for which ventilation was required for four days. After extubation, he did not utter words for three weeks, but only whined and cried with a soft voice, especially when stimulated to speak. Lower cranial nerves were intact except for mild lingual weakness. Complex movements of the mouth were severely impaired (Table 2.2.). After three weeks he suddenly started to produce monosyllabic words and within a few days he spoke in sentences of considerable length. Because of the language barrier, the speech could not be properly evaluated. However, the Dutch-speaking relatives described the language as fairly intelligible but slightly flaccid. During the mute phase, MRI showed quadriventricular hydrocephalus and a hyperintense contrast-enhancing lesion in the mesencephalic and pontine tegmentum, more so on the left than on the right side (Figure 2.3A). Measurements of intracranial pressure showed a low-pressure hydrocephalus. MRI two weeks after recurrence of speech showed that the hydrocephalus was diminished and the hyperintense lesion had disappeared (Figure 2.3B). When speech recurred, ataxia was still severe.

2.4. Results

The clinical data of the patients are summarized in Table 2.1. In the present series, none of the children was dysarthric before surgery. Five of the 15 children – all with a tumor lining the fourth ventricle – developed MSD after surgery. They became mute after one to two days with normal speech. None of the children with a left or right cerebellar hemisphere tumor developed dysarthria or MSD, even when the superior paravermal cortical region was clearly involved in the process (patients A1, A3, A5, and A7).

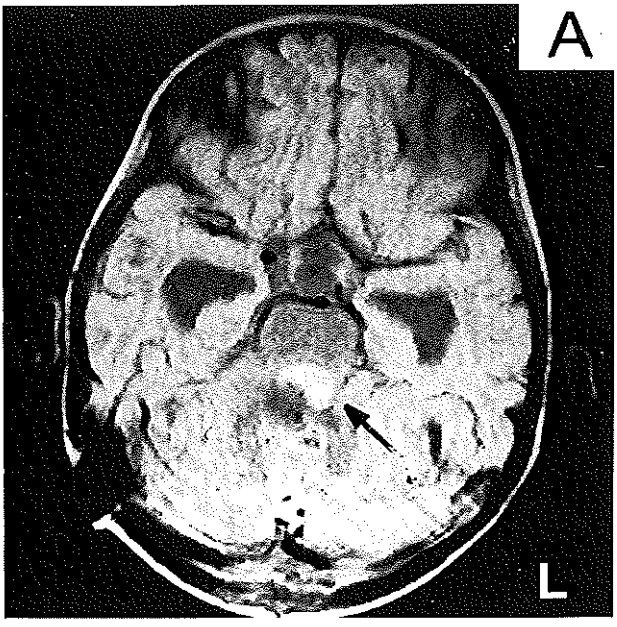
These patients all had astrocytomas, which may also cause MSD [4,6,8,9,12,14]. In all MSD children, we investigated the speech musculature. No paresis of bulbar musculature was found in group A children. Patients B1 and B2 showed pareses of bulbar nerves. Only a mild weakness of the tongue was found in all MSD children, but complex movements of the speech musculature – for example, alternating movements of the tongue – were disturbed during the mute phase in all the children.

The length of the mute phase varied from three to eight weeks. In patient C4, treatment with high-dosage bromocriptine, a dopamine agonist, did not influence recovery. The recurrence of speech, even from the point of view of the parents, was unexpected in all the children. When they regained speech they were all severely dysarthric, but speech recovered to almost normal surprisingly fast in one to five weeks. In four of the five children with MSD, speech could be studied in detail. Only patient C2 had one mild ataxic speech characteristic. Recovery of dysarthria to normal speech seems to be related to recovery of complex movements of mouth and tongue.

In all three groups, the sizes of the tumors varied considerably. The largest tumors were not present exclusively in the MSD group. In the five MSD children, the tumor involved the inferior part of the vermis cerebelli (Figure 2.2.) with exophytic growth into the fourth ventricle. In patients C2, C3, C4, and C5, tumor completely filled the fourth ventricle and adhered to the floor of the fourth ventricle. Tumor growth through the inferior cerebellar peduncle onto the ventral surface of the brainstem was present in patients A8, B1, and C5, all with ependymomas.
Four of 10 children from groups A and B needed a VP shunt at presentation. In contrast, in all MSD children, a major hydrocephalus was present that needed shunting prior to tumor surgery ($p < 0.05$, Fisher's exact test). Patient C5 had a quadriventricular normal-pressure hydrocephalus after surgery, and patients C3 and A8 had a shunt infection and needed shunt revision after surgery.
In three of the five MSD children, MRI was performed during the mute phase. In patients C3 and C4, MRI six and four weeks postsurgery revealed no abnormalities except for the surgical cerebellar lesion. In patient C5, MRI two weeks after surgery revealed edema in the pontomesencephalic tegmentum involving the crossing fibers of the superior cerebellar peduncles (Figure 2.3A). This edema had disappeared three weeks later, i.e. two weeks after recurrence of speech (Figure 2.3B). In contrast, after infarction of the ventral medulla oblongata in patient B2, no MSD occurred.

2.5. Discussion

We found that an isolated lesion of the cerebellar structures caused by removal of a hemisphere tumor was not sufficient to produce MSD. An additional ventricular localization of tumor and adherence to the dorsal brainstem were necessary risk factors [7,10,12], suggesting that brainstem dysfunction is important in the etiology of MSD. The obligatory postsurgical occurrence of MSD suggests that trauma is of importance in its evolution, possibly due to the more intense manipulation of the brainstem, when part of the tumor situated in the fourth ventricle or adherence to the dorsal brainstem is removed. Because of the transient nature of the syndrome, Ferrante et al. [7] suggested ischemia and possibly edema as possible mechanisms. Edema in the pontine tegmentum in patient C5 during the mute phase supports this assumption. The frequent occurrence of pyramidal and eye movement signs in MSD patients supports brainstem dysfunction. Neither exclusive cerebellar or brainstem lesions or hydrocephalus alone, but rather a complex interaction of these variables, causes MSD.



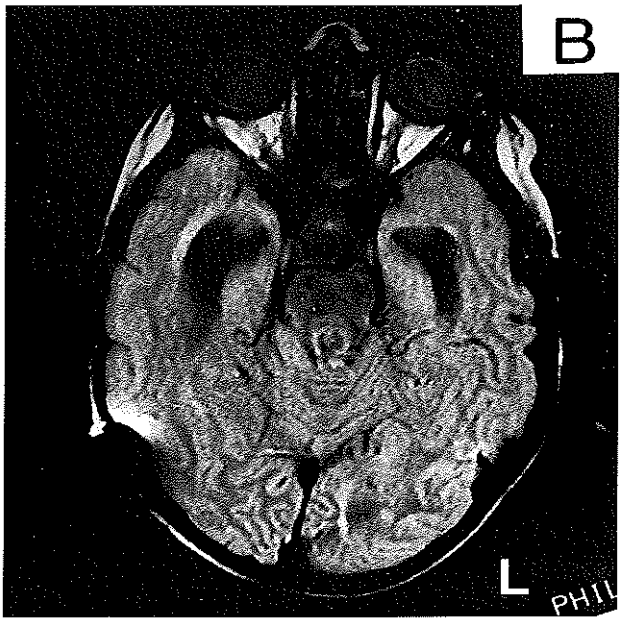


Figure 2.3. T1-weighted MRI of patient C5, 2 weeks (A) and 5 weeks (B) after resection of an ependymoma.
Note hyperintensive lesion (L>R) in pontine tegmentum in (A) (arrow) which is no longer present 3 weeks later (B).

Unlike adults [20-22], children do not suffer speech disturbance caused by damage to the superior paravermal cortical region, as patients A1, A3, A5, and A7 demonstrate. In these patients, whose CTs after surgery clearly showed either left- or right-sided damage to these regions, speech remained normal. Despite their normal speech, some of the children in group A were severely ataxic (patients A3 and A7), demonstrating the lack of strict relation between ataxia and speech disturbance [23].

In view of the multifactorial origin of MSD, it is not expected that a 'pure' cerebellar dysarthria will occur. The four patients of group C had a variety of speech characteristics. The most specific features of ataxic speech according to Darley et al. [23], which are *excess and equal stress* and *irregular articulatory break down,* were met in none of those four patients. Only one (patient C2) had a specific feature of cerebellar speech (alternating loudness) according to Kluin et al. [24]. The observed dysarthric features do not fit into any other cluster of deviant dimensions considered specific for a certain type of dysarthria [23].

In patient B2, extensive infarction of the ventral medulla occurred without MSD. Because of the possibility of dysfunction of the ascending mesencephalofrontal fibers originating from the A9 and A10 dopaminergic cell groups causing MSD [26-28], we administered bromocriptine to patient C4. The drug had no effect on the MSD.The above observation as well as the finding of intact bulbar cranial nerves in patients who developed MSD suggest a localization of the brainstem dysfunction rostral to the medulla oblongata and caudal to the mesencephalon.

Summarizing our data and those in the literature, we conclude that MSD is a childhood syndrome with the following features: (1) occurrence between ages two and 10 years; (2) transient mutism of variable duration, followed by dysarthria that recovers quickly and completely; (3) bilateral dysfunction of the dentatothalamic fiber bundles or their cells of origin after the surgical intervention in the posterior fossa as the proximate cause; and (4) a spectrum of neurological manifestations, varying in severity, as a frequent accompaniment.

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3

Update of the patient series, presented in chapter 2

Abstract
We present the data of 29 new referrals for posterior fossa tumor resection, of whom seven children suffered from MSD. As in the previous chapter (Chapter 2) we focus on the features of the mutism and dysarthria. Three MSD patients are described in detail, four patients were excluded from further analysis because of their age. The observations confirm previous findings (Chapter 2) with respect to the behavioral features during the mute phase and to the dysarthric speech features.

3.1. Introduction

In Chapter 2, published by Van Dongen et al. [1], we presented a consecutive series of 15 children who had undergone posterior fossa tumor resection between 1990 – 1993 (Group 1). Postoperatively five patients suffered from mutism and subsequent dysarthria (MSD) (Group 1C) and two children had mild speech problems (Group 1B).
We here present the data of 29 new referrals for posterior fossa tumor resection in the period 1994 – 1997 (Group 2). By adding these new cases we aim to further analyze the features of MSD. We compare the results with recently published cases and review studies [2-8].

3.2. Methods

In 1994-1997, 29 children (Group 2) with a posterior fossa tumor were surgically treated. All children had had a normal psychomotor development. Before admission no speech problems were noted. Speech was analyzed according to the same method [1]. We divided the patients of Group 2 as in Group 1: Group 2A (N=22) had no speech abnormalities after surgery. We did not observe the occurrence of speech abnormalities immediately following the operation, as was the case in two children in Group 1 (Group 1B). Group 2C (N=7) suffered from MSD.
The characteristics of the groups (Group 1A 1B, and 1C) and Group 2 (Group 2A and 2C) are summarized in Table 3.1.
Regarding the aims of the present study, we excluded four patients from further analysis: three patients were too young for proper speech evaluation (three-years-old) and one (17-years-old) was considered too old for inclusion in a pediatric group.

3.3. Patients

Patient C6. This 12-year-old boy was admitted because of clumsiness, frequent headaches, and vomiting during the preceding 12 months. Since four months he complained of diplopia and there was a head tilt. On examination, there was gait disturbance, bilateral papilledema, nystagmus and saccadic eye movements. MRI (Figure 3.1.A,a) showed a posterior fossa tumor and an associated hydrocephalus. A VP-shunt was inserted and four days later a medulloblastoma extending from the vermis into the fourth ventricle was removed. During the operation the arteria cerebelli posterior inferior had to be sacrificed. After the operation he was tetraparalytic, there was a paresis of the right facial nerve and he did not communicate. Two weeks after surgery he could be extubated. He had an epileptic insult for which he was treated with carbamazepine. One month postsurgery his clinical condition worsened. A repeat CT-scan did not disclose the cause of this deterioration. Chemotherapy was started, but this had no effect on his condition. Throughout the mute phase which lasted for five months, he was drowsy and hardly testable. On requests, he occasionally made correct articulatory voiceless movements.

Table 3.1. Data of children, who underwent posterior fossa tumor resection: Group 1 referred 1990-1993 (Chapter 2) [1] and Group 2 referred 1993-1997 (this chapter).

Patient groups	Group 1	Group 2
A. No speech problems		
Number	8	22
Mean age	6.6 yrs	8.1 yrs
Age range	2-12 yrs	3-17 yrs
Tumor type		
Astrocytoma	7	11
Medulloblastoma	-	8
Ependymoma	1	1
Hemangioblastoma	-	2
B. Mild dysarthria		
Number	2	-
Mean age	7 yrs	-
Age range	4-10 yrs	-
Tumor type		
Ependymoma	1	-
Meningioma	1	-
C. Mutism and subsequent dysarthria		
Number	5	7
Mean age	6.2 yrs	8.1 yrs
Age range	4-8 yrs	3-17 yrs
Tumor type		
Medulloblastoma	4	5
Astrocytoma	-	2
Ependymoma	1	-

Orofacial movements were impossible except for one successful performance of tongue protrusion. When speech recurred there was a moderate bilateral paresis, more pronounced in the arms than in the legs.

Recovery of speech Unexpectedly, speech recurred after five months, but the quality of speech was difficult to analyze, as he could only repeat some single words and a three-word-sentence. Three months later speech was only midly impaired. Voice was hypernasal {2} and slightly harsh {1}. Speech rate was slow {2}, but there were only mild articulatory problems, mainly consonant distorsion {1}.

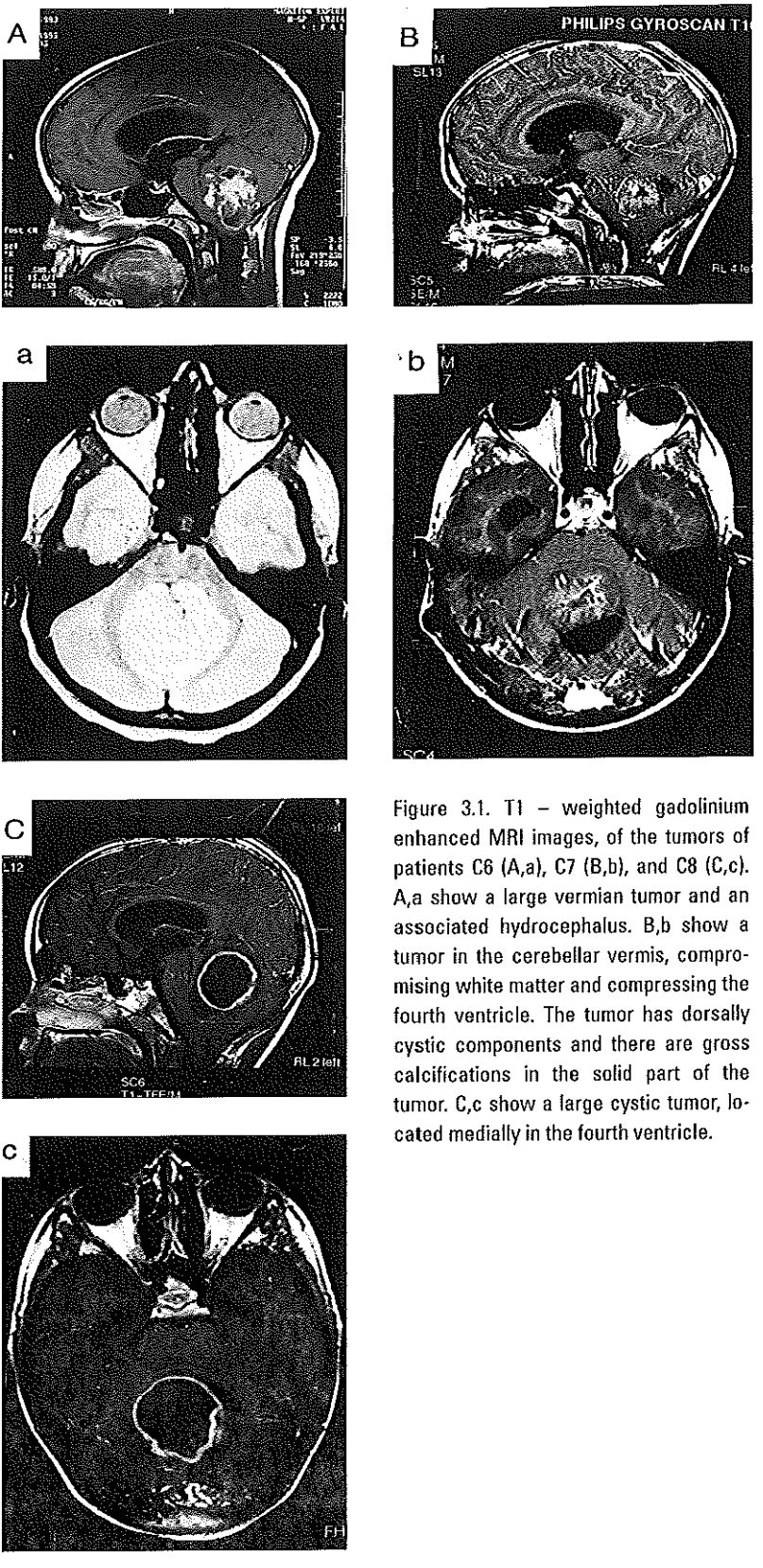

Figure 3.1. T1 – weighted gadolinium enhanced MRI images, of the tumors of patients C6 (A,a), C7 (B,b), and C8 (C,c). A,a show a large vermian tumor and an associated hydrocephalus. B,b show a tumor in the cerebellar vermis, compromising white matter and compressing the fourth ventricle. The tumor has dorsally cystic components and there are gross calcifications in the solid part of the tumor. C,c show a large cystic tumor, located medially in the fourth ventricle.

Patient C7. This 12-year-old boy had been referred with progressive diminution of vision, nausea, vomiting, and headache. His gait was spastic, there was diplopia and diminished vision. MRI (Figure 3.1.B,b) showed a tumor in the cerebellar vermis, compromising cerebellar white matter and compressing the fourth ventricle. The tumor had dorsally cystic components and there were gross calcifications in the solid part of the tumor (Figure 3.1.b). Cisterns were obliterated. A VP-shunt was inserted and three days later a medulloblastoma was subtotally resected. Postoperatively there was a paresis of the left arm, and a further deterioration of vision. On the second postoperative day he was drowsy, but he said 'yes' and 'papa'. On the following days he opened his eyes when addressed and responded by squeezing the fingers of the examiner, but he did not speak. One week postoperatively he was apathetic and kept his eyes closed. He could not perform orofacial movements except for tongue protrusion and eye opening. In the following weeks he became more alert, the function of the left arm improved, but his vision was still severely impaired. Language comprehension seemed intact, as he correctly responded to requests as 'point to your ear' and 'lift your arm'. Three weeks postperatively he could perform simple orofacial movements, such as lateral tongue movements, but on requests for more complex movements he opened his mouth widely and stereotypically. He did not phonate, except when laughing. Speech recurred four weeks after the operation.
Recovery of speech. Three days after he had started to speak the severe dysarthria was characterized by the following features: hypernasality {3} and hoarse voice {3}, imprecise consonants {3}, and distorted vowels {3}. Speech rate was very slow {3}. One week later hypernasality had lessened {1}, and articulation had improved to some extent. Again after one week consonants were still imprecise {2} and vowels distorted {2}, contributing to a still slow speech rate {2}, and his voice was still slightly hoarse {1}.

Patient C8. This seven-year-old boy had a four weeks history of progressive headaches, vomiting and unsteady gait. At admission there was a left-sided pyramidal syndrome and left-sided ataxia. MRI showed a large cystic tumor, located medially in the fourth ventricle (Figure 3.1. C, c). This pilocytic astrocytoma was removed in toto and a frontal drain was placed. Postoperatively there were no new neurological deficits. On the first postoperative day he spoke with a whining voice. He was reluctant to speak, but we could not detect any abnormal speech features. Moreover, tongue protrusion and some complex orofacial movements were performed correctly. However, on the morning of the third postoperative day he whined and turned away, when addressed. He did not speak.
Recovery of speech. In the afternoon of the same day he spoke again. Speech rate was slightly slow {1}. Four days later his speech was not dysarthric except for a slightly low pitch {1}. He could repeat long sentences. Complex orofacial movements were performed correctly.

3.4. Results

In these three children who developed MSD postoperatively, VP shunting preoperatively was required in two cases, a frontal drain was placed during the operation in the third case. Tumors extended into the fourth ventricle. Tumor type in Cases C6 and C7 was a medulloblastoma and in Case C8 a pilocytic astrocytoma. The length of the mute phase varied from half-a-day to five months. During the mute phase we observed impairments of voluntary complex orofacial movements. Table 3.2. presents data on the function of cranial nerves and quality of orofacial movements, shortly after the recurrence of speech.

Table 3.2. Results of neurological examination of lower cranial nerves during the initial stage of dysarthria.

Symptom (cranial nerve)	Patient C6	Patient C7	Patient C8
Trigeminal weakness (V m)	N	N	N
Sensation (V s)	N	N	N
Facial weakness (VII R/L)	+/N	N	N
Pharyngeal weakness (IX)	+	N	N
Lingual weakness (XII)			
Tongue protrusion	N	N	N
R/L movements	N	N	N
Complex movements			
Alternating tongue movements	+	+	N
Clicking	N	+	N
Chewing	N	N	N
Swallowing	N	N	N
Blowing	N	N	n.a.
Coughing	N	N	n.a.

m Motor branch
s Sensory branch
R Right
L Left
N No abnormalities
+ Mildly abnormal
n.a not assessed

Bulbar pareses were absent except for a right peripheral facial nerve paresis in C6. The prominent features of the severe dysarthria were slow rate, and articulatory impairment in cases C6 and C7. The dysarthria recovered slowly in C6, as there was still slow speech rate and hoarse voice three months after the onset of dysarthric speech.

3.5. Discussion

The localization of the tumor in the cerebellar vermis, its extension into the fourth ventricle, and tumor type in two cases (C6, C7) were in accordance with our previous study (Chapter 2) [1] and more recent studies [2,7,8]. The findings in the present group of patients confirm earlier findings regarding the pathogenesis of MSD as reported in the previous study (Chapter 2) [1].

The duration of the mute phase varied considerably. Patient C8 was mute for half a day with a slight speech impairment in the following week. His behavior was similar to which we had observed in Case C4 (Group 1) – whining and turning away when addressed –, which was in contrast with his speech and behavior on the first postoperative day. Speech was slightly dysarthric. We therefore labeled this clinical picture MSD. The duration of the mutism was extremely long in C6, compared to recent reviews, reporting a duration of four days to four months [8]. This long duration was regarded as a gloomy prognosis for speech recurrence and therefore the onset of speech after five months was quite unexpected.
During the mute phase, orofacial movements could not be made on request, but were restored shortly before or after speech onset. It is worth mentioning that we observed normal orofacial movements on the first postoperative day in Case C8. Recent studies [5,6] have proposed the 'apraxic' nature of mutism, which had been suggested earlier by Ferrante et al. [9]. In this study, the loss of verbal expression was attributed to a 'language apraxia', 'an inability to coordinate the movements of the organs of phonation (vocal cords, tongue, facial and respiratory muscles)'[9]. Dailey et al. [5] observed difficulties 'coordinating the oral and pharyngeal musculature as manifested by postoperative drooling and inability to swallow', labeled 'oropharyngeal apraxia'. Pollack et al. [6] also suggested that mutism resulted from an inability to initiate complex volitional movements. In contrast, Crutchfield et al. [3] reported that after mutism, words and sentences immediately return, which points to an 'inhibition' rather than an apraxia. Al-Jarallah et al. [2] found no evidence that the mutism was secondary to paralysis or apraxia of speech musculature. In light of these controversial hypotheses, we performed a seperate study on the role of voluntary movements during mutism and shortly after the onset of speech (Chapter 5) [10].
Analysis of the dysarthric speech features in the presented cases further demonstrated, that scanning speech and irregular articulatory breakdown – the hallmarks of ataxic dysarthria in adults [11,12] – did neither occur in the group described in Chapter 2 nor in the present group. These observations lead to a systematic analysis of speech features in children with MSD. These data are reported in Chapter 7.

We conclude that MSD constitutes a spectrum of variable duration and severity. The very short period of speechlessness in Case C8 may be caused by discrete neurological events, which exact nature remained unknown. The extremely long mute phase in Case C6 was caused by multiple factors, similar to the cases, described in Chapter 2 [1]. The presented cases suggest, that the restoration of complex orofacial movements is crucial for the onset of dysarthric speech (Chapter 5)[10].

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The many faces of acquired neurological mutism in childhood

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4

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Abstract
Acquired neurological mutism in childhood is a complex phenomenon occurring in various neurological conditions with different etiologies. We illustrate its clinical heterogeneity as reflected in a wide range of concomitant behavioral features by presenting four children with acquired neurological mutism. Neuropsychological examination revealed differential patterns of defective or preserved phonation, orofacial movements, communicative behavior, and linguistic functions. We propose that detailed neuropsychological analysis contributes to descriptions of the evolution of the speech impairment beyond the mute phase and the long-term disability. A framework for the clinical evaluation of mute children is therefore presented.

4.1. Introduction

Patients are diagnosed with neurological mutism when an organic lesion can reasonably be held responsible for the absence of verbal output or when the mutism is associated with other deficits suggestive of dysfunction of the central nervous system (CNS) [1]. Acquired neurological childhood mutism (ANCM) may be caused by damage in different brain regions in the context of diverse etiologies (Table 4.1.) [2-21].

Table 4.1. Illustrative case studies of acquired neurological childhood mutism.

Reference	Localization	Etiology
[2]	Left cerebral hemisphere	Traumatic
[3]	Left cerebral hemisphere	Vascular
[4]	Left cerebral hemisphere	Traumatic
[5]	Left cerebral hemisphere	Traumatic
[6]	Right cerebral hemisphere	Vascular
[7]	Perisylvian areas	Epileptic
[8]	Perisylvian areas	Epileptic
[9]	Opercula	Infectious
[10]	Opercula	Epileptic
[11]	Opercula	Vascular
[2]	Diffuse	Traumatic
[12]	Cortical/subcortical pathology	Infectious
[13]	Thalamus	Traumatic
[14]	Third ventricle	Tumor
[15]	Basal ganglia	Infectious
[16]	Brainstem	Tumor surgery
[17]	Brainstem	Tumor
[19-21]	Cerebellum	Tumor surgery

ANCM may therefore be associated with a wide range of neurological and neuropsychological deficits such as cranial nerve paresis, absence of phonation, disturbed simple and complex orofacial movements, absence of communicative urge, or loss of comprehension of spoken language.

Pediatric studies usually mention the presence of ANCM in case studies as a sign of a specific disease, but associated behavioral features have not been analyzed in detail. Therefore, the possible prognostic value of such features for recovery from mutism or for long term sequelae is not known. We wished a) to illustrate the heterogeneity of the neuropsychological disorders that contribute to the many faces of ANCM and b) to provide a framework for the clinical evaluation of mute children.

Table 4.2. Behavioral features during the mute phase of ANCM.

Feature Age of onset Time post onset	Case 1 11 yrs 4 months	Case 2 5 yrs 6 months	Case 3 9 yrs 1 month	Case 4 5 yrs 2 weeks
Alertness	'wakeful'	attentive and cooperative when tested.	attention could be drawn only by touching and stimulating.	attentive and alert to daily activities and when tested.
Motor behavior	tetraplegia left >right; 'motionless' except for slow eye movements and head turning when addressed	right-sided central facial nerve paresis and hemiplegia	no abnormalities except for a previously existing ataxia.	severe truncal and limb ataxia.
Orofacial movements	orofacial movements could not be elicited; drinks and swallows with help; smacks his lips; no facial expression.	normal chewing, swallowing; simple and complex oral movements impaired;vivid emotional expression; incidentally unphonated articulatory movements.	normal chewing, drinking and swallowing; little facial expression.	eats and drinks greedily; simple orofacial movements possible; complex learned movements impossible; vivid emotional expression.
Phonation	completely silent.	emotional vocalisation: giggling, growling.	completely silent.	whining and loud sobs; normally phonated laughing.
Communication	'promise of speech'[14]	good eye contact; nodding assent, shaking head; contextually adequate gestures.	reacts cooperatively upon gestures; no initiative to communication.	attentive listener.

Language	absent for all modalities	understands simple commands within context.	neglect of (repetitive) verbal and nonverbal auditory stimuli.	good language comprehension.
Mental activity	'mindless'[14]	limited attentional span: concentrated for half an hour; bradyfrenous; sorting of three forms by trial and error; nonverbal IQ at 4-yrs level.	aloof; no purposeful actions.	good performance at complex form sorting task; anticipatory and cooperative when tested.
Emotions	'mindless'[14]	cheerful most of the time; strong emotional reactions in response to environmental changes; impulsive.	no sign of emotional awareness.	contextually adequate expression of internal state such as frustration, pain, pleasure.

4.2. Case reports

We describe four children with ANCM. Cases 1, 2, and 4 previously had normal psychomotor and language development. Case 3 had developmental motor problems, but normal speech, language, and scholastic ability. All children were examined identically and videotaped on several occasions. Table 4.2. describes the characteristic neurobehavioral features in each patient to illustrate distinct clinical presentations of ANCM.

Case 1. An 11-year-old boy sustained severe head injury. On his hospital admission the Glasgow Coma Scale (GCS) was E1M1V1.
CT scan disclosed hemorrhages in the left frontoparietal region and in the right insula. Epileptic seizures occurred frequently. EEG showed slow background activity with epileptic foci in the right temporo-parieto-occipital and in the left frontal regions. In the next three weeks he developed spastic quadriplegia. He could suck and swallow and could eat when fed. His eyes followed moving objects and persons approaching him. He made no attempts to speak although he 'seemed to give promise of speech'. He could not be stimulated to communicate in any other way. He did not show facial emotional expression and did not phonate in emotional context. Four months after the accident, in a videotaped session he was still akinetic and mute (Table 4.2.).
The mutism lasted for approximately one year. Fifteen months after the accident he was wheelchair bound. He was cooperative and communicative. He understood verbal requests but exhibited marked mental slowness. His severe dysarthria was characterized by slow speech rate and breathiness to such a degree that he stopped speaking after five minutes conversation. Grammatical form of his utterances and wordfinding were unimpaired.

Case 2. A five-year-old girl was injured in a car accident. On admission to the intensive care she was comatose (GCS: E1M5V1). CT scan demonstrated fractures of the left parietal and temporal bones, multiple areas of contusion in the left hemisphere, obliterated basal cisterns and shift of the midline structures towards the right side. Intracranial pressure in the first hours was increased to 50-60 mm Hg, and she was intubated and artificially hyperventilated. Neurological examination revealed a right flaccid hemiparesis, including a right central facial nerve paresis and a bilateral abducens nerve paralysis. Sixteen days after onset she was extubated. She became more responsive to her environment: she reached for objects and waved goodbye. She did not speak but phonated in emotional context.
In the next six weeks her mobility improved rapidly. A session was videotaped six months after the accident (Table 4.2.).
Fourteen months after the trauma she produced the first word, 'Mama' but with articulatory problems. Three years after the accident she is still severely aphasic: language comprehension is severely impaired, and her verbal output shows articulatory problems and loss of syntax.

Case 3. The medical history of Case 3 is complex: he had been examined at the age of three years for delay of motor development. Neurological examination revealed

slight ataxia of all extremities. CT – scan did not demonstrate cerebellar pathology or other abnormalities, and the etiology remained unknown. Despite his delayed motor development, his speech and language developed normally.
At the age of five years, he fell from his tricycle and was admitted to the hospital with brain concussion, from which he recovered with no additional deficit. His school performance was age-appropriate.
In the acute phase, at the age of nine years, he fell again from his bicycle and complained of dizziness and blurred vision. In the next hours consciousness deteriorated. Neurological examination disclosed a restless, confusional state (GCS: E3M4V3). There were no signs of meningeal irritation or increased intracranial pressure. Pupils were of equal size and reacted to light. One day after hospital admission, he was still restless: he groaned to painful stimuli and uttered only a few words. There were bilateral Babinski responses. CT scan demonstrated narrow ventricles but no focal abnormalities. The course was complicated by increased body temperature. A second CT scan and cerebrospinal fluid were normal. His verbal output deteriorated within one day: he vocalized unrecognizable sounds (GCS: E3M3V2), after which he became completely mute.
One month after the trauma we analyzed his behavioral features (Table 4.2.). Neuropsychological examination initially revealed a neglect for all verbal and nonverbal auditory stimuli: cortical deafness. He did not startle to sounds and behaved as though he were totally deaf. He did not appear to be concerned about his inability to hear. Ear, nose, and throat examination and brainstem auditory evoked potentials were normal.
Five weeks after the trauma occurred, he became aware of spoken language and sounds but was unable to recognize their meaning: auditory agnosia. However, he was able to write the names of visually presented objects. Two weeks later he would correctly identify 50% of auditorily presented sounds, such as musical instruments. He still demonstrated auditory language problems – verbal auditory agnosia – to such a degree that he could not point to objects for which the names were presented auditorily, whereas he responded adequately to written words. Eleven weeks after the trauma his language comprehension had returned to normal. Moreover, his language production had gradually improved. He spoke with a whispering voice and was able to repeat long sentences. He had slight wordfinding difficulties.
One year after the accident, he had regained his premorbid level of functioning except for mild wordfinding difficulties. Five years after this episode, he has no language problems and his school performance is at age-appropriate level.

Case 4. A five-year-old boy was admitted with early morning vomiting, diplopia, and staggering gait. Neurological examination demonstrated bilateral papilledema, a vertical skew deviation and a slight ataxia of the left arm and leg were found. CT and MRI scan revealed a tumor of the cerebellar vermis with some small calcifications and cystic components filling the fourth ventricle and associated hydrocephalus. A ventriculoperitoneal shunt was inserted. In a second surgical session a medulloblastoma was completely removed macroscopically. Postoperatively, he had severe ataxia of trunk and limbs and he was extremely apathetic. He spoke only a few words for two days, after which he became mute.

The mutism lasted five weeks. Emotional phonation (crying, whining, laughing) was intact throughout the mute phase. Coordination of movements during eating and swallowing were intact. He could lick off his lips while eating but voluntary orofacial movements could not be initiated on request despite intact language comprehension. Later on, simple orofacial movements such as eye closure, tongue protrusion and lateral tongue movements were performed adequately. Complex learned voluntary movements such as whistling, blowing, imitating an angry face and blowing a kiss could be performed adequately soon before the mutism disappeared. Subsequently, he was dysarthric: he spoke single words with slow speech rate and a strained strangled voice. Three months postoperatively, his spontaneous speech was limited, but he was able to repeat sentences slowly. Four months after the tumor resection, speech was normal except for a slightly slow rate. (Further details are reported by Van Dongen et al.(Case C4) [21])

4.3. Summary of the cases

In accordance with the original description by Cairns [14] and later studies [12,13,22], *Case 1* is a typical example of the *akinetic mutism:* the patient is immobile, completely silent and nonresponsive. Therefore the mutism in this context is nonselective and part of a global lack of responsiveness. In this clinical context the associated cognitive and behavioral features cannot be assessed as was possible in the other cases. In Case 1, the probable speech disorder associated with the mutism was anarthria since severe dysarthria followed the mute phase. Aphasia could not be excluded but was unlikely since the boy did not show manifest impairments of language comprehension after the mute phase.

In *Case 2*, it was evident that the mutism was an initial phase of *severe aphasia*, since evaluation during the mute phase demonstrated severe impairment of language comprehension. The patient was eager to communicate, but had limited ability to convey information by alternative means such as gestures. This severe aphasia has influenced the long duration of the mutism. Three years after the onset of mutism the girl is still severely aphasic.

Mutism has a high prevalence in childhood aphasia [3,4] in which disorders of language comprehension are common [23]. Case 2 shows remarkable resemblance to the case described by Jordan [24] with incomplete recovery over a protracted period after the 'return from silence'.

In *Case 3*, the mutism occurred in the context of a severe *cortical deafness* i.e. 'apparent unawareness of auditory stimuli in the absence of peripheral or brainstem damage'[25]. Subsequently he manifested an auditory agnosia: he was aware of auditory input but unable to recognize its meaning. Speech returned when he became able to understand nonverbal sounds and language comprehension improved. After one year, he exhibited only mild wordfinding problems.

Case 4 presented with a rather pure *mutism after resection of a cerebellar tumor:* during the mute phase, he was able to phonate, he understood language at age-appropriate level as was demonstrated by adequate performances on receptive language tasks and he performed well on nonverbal cognitive tasks. However, initiation of voluntary

complex movements was significantly impaired. The onset of dysarthic speech coincided with the recovery of voluntary complex orofacial movements. Therefore we hypothesize that the mutism was due to a loss of the ability to initiate and coordinate complex orofacial movements. Such a clinical picture resembles other cases with mutism due to posterior fossa surgery [18-21].
The most relevant data of the four Cases are summarized in Table 4.3.

Table 4.3. Summary of behavioral analyses.

Parameter	Case 1 Akinetic mutism	Case 2 Aphasic mutism	Case 3 Cortical deafness	Case 4 Posterior fossa mutism
Alertness	–	+	+/-	+
Cranial Nerves	+	right-sided VII, bilateral VI	+	+
Orofacial movements	?	+/-	+	+/-
Phonation	–	+	–	+
Communicative intent	–	+	–	+
Language comprehension	?	+/-	–	+

\+ intact; - severely impaired
+/- moderately impaired; ? could not be assessed

4.4. Discussion

ANCM is a complex phenomenon occurring in neurological disease in childhood. The underlying mechanisms are various, resulting from damage anywhere between the cerebral cortex and the peripheral speech apparatus. The pediatric literature on ANCM consists of case studies describing the occurrence of mutism and presenting occasional definitions in a specific clinical context. Our cases were examined and presented in a standard fashion to illustrate the many faces of ANCM and to distinguish various forms of mutism as a manifestation of distinct underlying mechanisms. On the basis of our analyses and other studies, we discriminate between the following causes: deficient initiation of cognition and speech, auditory agnosia and aphasia, and dysarthria.
Deficient initiation of cognition and speech. Akinetic mutism (AM) is probably the type of mutism best known to clinicians. It is a neurobehavioral condition characterized by deficient initiation or activation of behavior and cognition. Case 1 fits the criteria for AM, which are distinct from those of the vegetative state (VS) [22]. Case 1 followed requests to suck and swallow, whereas in VS behavior responses consist of reflexes only. Visual tracking, crucial to the diagnosis of AM, is more sustained in AM

than in VS, in which visual tracking, if present, extinguishes rapidly. In addition, in AM the facial expression shows 'promise of speech' [14] which is absent in VS, in which signs of communication with the environment are absent [22]. In most cases of AM, a minimal degree of movement and / or speech can be elicited depending on the nature and intensity of stimulation provided [22]. We could elicit no emotional phonation or speech in Case 1.

Auditory agnosia and aphasia. Case 3 manifested cortical deafness resolving into auditory agnosia, which suggests that in this child the loss of meaningful auditory input precluded verbal output. This is in contrast to conditions in adults with cortical deafness, who retain the ability to produce speech [25].

Mutism has a high prevalence in acquired childhood aphasia (ACA) [3,4]. It may be related to a language comprehension disorder as in Case 2. After the 14 months mute phase, her verbal output was severely aphasic and language comprehension was impaired. In contrast to earlier notions, a more recent description of ACA states that impaired auditory comprehension is detected in almost all aphasic children if appropriate tests are administered and may range from mild difficulties to total loss of receptive communication [23].

In acquired aphasia with seizure disorders – Landau Kleffner syndrome – the severe disruption of language comprehension (verbal auditory agnosia) may also result in loss of verbal output (mutism) [26]. This clinical picture has been described only in children. In adults, 'aphasic' mutism is usually transient, even in types of aphasia in which language comprehension is severely impaired [27]. However, mutism is not necessarily linked to language comprehension disorder. It may also be the manifestation of a severe articulation deficit with relatively preserved language comprehension, as may be the case in Broca's aphasia or verbal apraxia [5].

Mutism as an initial stage of ACA may be related to frontorolandic lesions [4] or subcortical lesions [3]. Furthermore, a language comprehension disorder may be a greater risk factor in childhood than in adulthood for mutism of long duration.

Dysarthria. Interference with any of the basic motor processes involved in speech production results in dysarthria. Anarthria, mutism of dysarthric nature, is the most severe manifestation. Damage, affecting motor subsystems may affect motor aspects of speech in various ways.

Bilateral damage to the anterior opercular region of the primary motor cortex causes loss of volitional control of muscles innervating lips, jaw, tongue, velum, and pharynx, rendering the patient mute with constant drooling. In contrast, involuntary movements (yawning, laughing) are preserved [9]. Patients are alert and have normal language comprehension. Mutism may be the main neurological sequela after structural damage to the anterior opercula. In contrast, in the opercular syndrome of epileptic origin, discharges in the opercular region cause only brief episodes of speech arrest [7,8].

Mutism may also occur in the context of an akinetic-rigid syndrome, as described by Pranzatelli et al. [15], who reported six children with acquired parkinsonism in whom mutism was associated with drooling and dysphagia in the context of such a syndrome. After appropriate medication, the mutism resolved into dysarthric speech.

Mutism after resection of a posterior fossa tumor has received much attention in the literature [18-21]. After an initially uneventful recovery from the operation, children

cease to speak. The children are alert and perform in age-adequate manner on nonlanguage tasks; their language comprehension is adequate [18-21]. The mute phase is of variable duration and is followed by severe dysarthria with good recovery. With respect to the pathophysiological mechanism, we previously observed [21] that in addition to the surgical lesion of cerebellar structures, ventricular localization of the tumor, adherence to the dorsal brainstem, and hydrocephalus at presentation were necessary risk factors. The motor impairment underlying the loss of speech is unknown. Observations in our patient (Case 4) suggest that mutism may be associated with the inability to initiate voluntary complex orofacial movements.
Frim and Ogilvy [16] reported mutism with subsequent cerebellar dysarthria after the surgical resection of a cavernous malformation of the right pons at the level of the middle cerebellar peduncle. Brainstem lesions may also produce locked-in syndrome, in which state patients are alert, cognitively aware of their environment, and capable of communication but incapable of speech or movement. The hallmark signs are quadriplegia and aphonia, caused by lesions involving the ventral pons [22].

A further distinction in types of mutism was made by Cummings et al. [28] and Von Cramon [29], who distinguish between the loss of emotional vocalization and the loss of propositional speech. They provide evidence that the limbic system is involved in phonatory expression of emotion such as laughing and crying, whereas descending neocortical systems subserve propositional speech and volitional facial movements. Von Cramon [29] followed adult patients with traumatic brain injuries during the mute phase and reported that 'complete' mutism was followed by a phase of nonverbal emotional vocalization (verbal mutism), signaling the recovery of midbrain function. In our cases, we could distinguish between complete mutism (Cases 1 and 3) and loss of propositional speech with preserved phonation in laughing, whining, and crying (verbal mutism) (Cases 2 and 4). These observations may add to the clarification of the neuropathological mechanism underlying mutism.

By identical examination of four mute children, we have illustrated that mutism is associated with various neurobehavioral features.
Our study may be considered an attempt to map the heterogeneity of ANCM and its underlying mechanisms. Detailed neuropsychological analysis as proposed in Appendix 1, is pertinent since the lack of speech itself provides no indication of the underlying mechanism. This analysis contributes to early statements as to the evolution of the speech / language problem and the expected longterm disability.

Appendix 1

Protocol for the evaluation of mute children.

1. **Cranial Nerves (lower)**
   Drinking, chewing, swallowing, coughing;

2. **Involuntary facial movements**
   Observation of laughing, yawning, facial expression;

3. **Simple orofacial movements**
   Eye closure, tongue protrusion, lateral tongue movements, licking off the lips, lateral jaw movements, showing teeth;

4. **Complex orofacial movements**
   Whistling, blowing, clicking of the tongue, showing an angry face, pretending to sense a nasty smell, blowing a kiss;

5. **Phonation**
   Absence or presence of phonation, emotional voiced utterances such as crying, whining, laughing, isolated voiced sounds;

6. **Auditory input**
   Localisation and identification of nonverbal sounds such as environmental noise, animal sounds, musical instruments;

7. **Language comprehension**
   Simple commands (close your eyes), complex commands, language comprehension tests (Peabody Picture Vocabulary Test, Test for Reception of Grammar (TROG), reading text);

8. **Alternative means of communication**
   Shaking, nodding, gesturing, writing;

9. **Nonverbal tasks**
   Making puzzles, sorting tasks, nonverbal intelligence tasks.

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Complex orofacial movements and the disappearance of cerebellar mutism

Report of five cases

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5

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Abstract
A syndrome of mutism and subsequent dysarthria (MSD) occurs frequently in children after resection of a cerebellar tumor. The role of orofacial and speech motor control in this syndrome has not been studied systematically. We examined simple and complex orofacial movements during the mute phase and shortly after onset of speech in five children with MSD. The recovery of complex orofacial movements coincided with the disappearance of the mutism.

5.1. Introduction

In children resection of a posterior fossa tumor may cause a syndrome of 'cerebellar mutism and subsequent dysarthria'(MSD) [1]. This syndrome has been described in approximately 75 children [1-18].
Mutism – complete loss of speech – occurs within a few hours to nine days after the resection of the tumor and may last one to 20 weeks. Speech recurs unexpectedly for parents and hospital staff. Initially speech is severely dysarthric but recovers rapidly.
A few studies refer to the quality of orofacial musculature during the mute phase. Most studies report sufficient function of the lower cranial nerves to such an extent that chewing and swallowing are intact. In contrast with these findings, Dailey et al. [15] report difficulty of coordination of oral and pharyngeal musculature during swallowing. In some studies orofacial musculature has been examined more fully: Rekate et al. [2], Dietze and Mickle [5] and Al-Jarallah et al. [12] observed normal movements of lips, tongue and palate. In an earlier study [8] we noticed that complex orofacial movements were impaired, but we did not analyze this impairment systematically. Murdoch and Hudson-Tennent [19] examined oromotor abilities, but their analysis took place at least one-and-a-half year after surgery after an extensive period of radiotherapy and chemotherapy. Thus, detailed analysis of simple and complex orofacial movements during the mute phase and at onset of speech has not been carried out. Therefore, the significance of these movements and of their recovery for the timing of speech onset is not known.

We followed five children who underwent resection of a posterior fossa tumor and subsequently suffered from MSD. We examined simple and complex orofacial movements during the mute phase and shortly after onset of speech.

5.2. Patients

Patient data are given in Table 5.1.. Preoperatively speech, language and motor development were normal in all children. Medulloblastoma outnumbers other tumor types which is in accordance with epidemiological data for this syndrome [1,16,17]. Tumors were located in the midline. In Case 2 the tumor also invaded the fourth ventricle. All patients suffered from ataxia of trunk and limbs postoperatively. Ventricular shunting for hydrocephalus was required in all patients prior to resection of the tumor. Postoperative complications were frequent. At the onset of speech all children were dysarthric[1].

1 For additional information on some of the subjects we refer to our previous studies: neurological findings in Cases 3, 4, and 5 were described by Van Dongen et al. [1] (Case C2, C3, and C4), speech features of Cases 3 and 4 were described by Catsman et al. [8] (Cases 2 and 3).

5.3. Methods

Assessments occurred at different intervals throughout the mute phase and after onset of speech (Figure 5.1.) within the limits imposed by the poor clinical condition of the children and intercurrent complications. For the examination of orofacial movements we closely followed the procedures of De Renzi et al. [20], Darley et al. [21], and Lehmkuhl et al. [22] for the testing of orofacial motor control in dysarthric and aphasic patients. We examined six SIMPLE OROFACIAL MOVEMENTS i.e. eye closure, tongue protrusion, lateral tongue movements, opening and closure of the jaw, lateral jaw movements, showing teeth; and nine COMPLEX OROFACIAL MOVEMENTS: chewing, blowing, coughing, whistling, clicking of the tongue, showing an angry face, pretending to sense a nasty smell, blowing a kiss, and teeth chattering. Performance was rated on a three-point scale: unimpaired (2), partial response or delayed response (1), no response (0). Maximal performance was 30 points. The children were verbally requested to make the orofacial movements. If this failed the child was encouraged to imitate. Communicative behavior and comprehension of spoken language were carefully observed. During the mute phase language comprehension was tested with items of the Test for the Reception of Grammar (TROG) [23] in Cases 2,4, and 5, as well as writing on dictation in Cases 1 and 4. Speech was rated according to the Mayo Clinic List of speech dimensions [24].

5.4. Case reports

Case 1. (See Table 5.1.) This 17-year-old boy had a two-months history of headache, dizziness, vomiting, and double vision. Neurological examination revealed papilledema and convergent strabismus. There was a slight ataxia of the right arm but gait was normal. Magnetic Resonance Imaging (MRI) – scan showed a large solid tumor in the midline with associated triventricular hydrocephalus. A ventriculo-peritoneal (VP)- shunt was inserted after which headache and double vision disappeared. Two weeks later the tumor was subtotally resected.

On postoperative day 1 the boy was alert and speech was adequate. In the course of the following day he developed a severe trunk and limb ataxia and orofacial dyskinesia was observed. He became mute. His behavior was characterized by a marked lack of spontaneity. He occasionally communicated by squeezing the hand of the examiner or pointing to objects. His language comprehension was normal. Moreover, he could write his own name and the names of presented objects. MRI – scan of the brain did show the surgical defect, but no other abnormalities. A single-photon emission computed tomography (SPECT) with ^{99m}Tc-HMPAO demonstrated a severe reduction of HMPAO uptake in the cerebral frontoparietal area.
Assessment of orofacial movements was started on postoperative day 13, four days before the onset of speech (Figure 5.1.). Facial expression was absent. He was able to lick his lips while eating and he could drink with little sips without choking. The mouth was almost continuously dyskinetic, predominantly the lips and the right

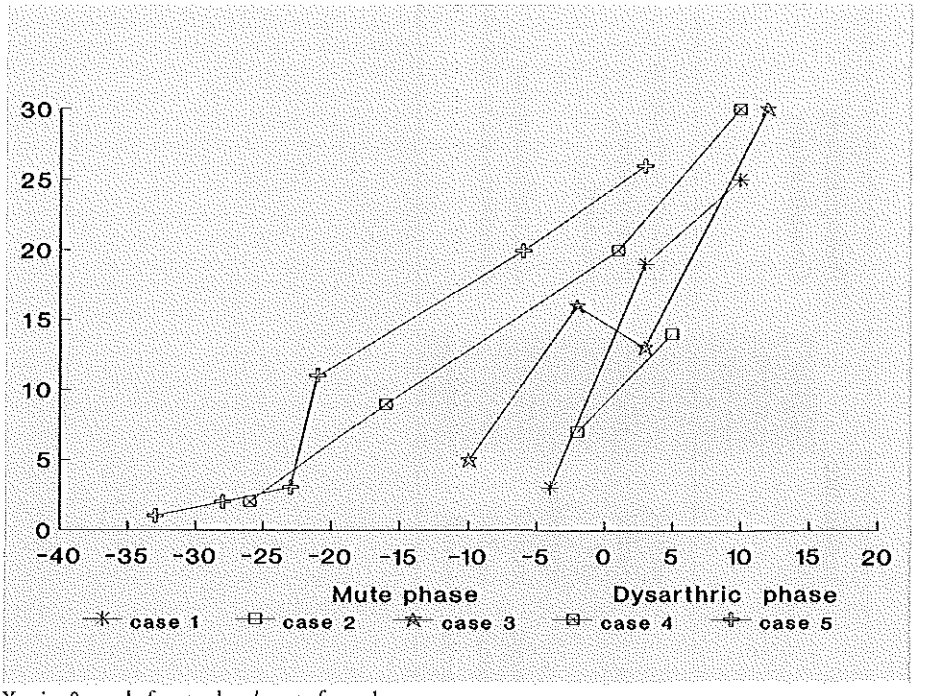
Table 5.1. Patient data.

Case	Sex/age	Tumor type	Lesion site	Lower cranial nerves	Postoperatively Complications	Onset of mutism (days po)	Onset of speech (days po)
1	M/17	astrocytoma	medial	normal	epidural haematoma	2	17
2	M/8	ependymoma	medial +IV ventricle	normal	shunt revision, infection	9	18
3	F/8	medulloblastoma	medial	VII, XII	epidural haematoma	2	60
4	M/8	medulloblastoma	medial	XII	pneumonia,shunt revision	1	60
5	M/5	medulloblastoma	medial	normal	-	1	36

M male
F female
po postoperatively.

corner of the mouth. Even when the mouth was at rest, simple or complex orofacial movements could not be performed except for eye closure. Four days after he had started to speak he could make all simple orofacial movements correctly and some complex movements. One week later almost all orofacial movements were carried out correctly.
At speech onset he whispered. In contrast with the verbal output of the other patients, his speech rate was normal and he produced long complex sentences. Speech improved within a week but voice volume was still reduced.

Figure 5.1. Performances on orofacial movement tasks.



X-axis: 0 = end of mute phase/onset of speech.

Case 2. At the age of five years a large ependymoma (grade II) of the fourth ventricle had been resected from which the boy recovered without complications. At the age of eight he was again admitted after a three week period of apathy, vomiting, loss of weight, dizziness, and unsteady gait (Table 5.1.). Neurological examination showed a first grade nystagmus to the left and a slight ataxia of the legs and broad based gait. The recurrent tumor was partially resected.
On the first postoperative day he had a mild trunk and limb ataxia. There was a first grade nystagmus, but no abnormalities of the lower cranial nerves. He was alert and he said that he wanted to go home. Shortly thereafter, the course was complicated by leakage of spinal fluid from the wound. This was treated by shunt revision and lumbar punctures. One week after the operation he became apathetic. He had severe

headaches with raised body temperature. He was treated as having a meningitis despite negative cultures from spinal fluid. On day 9 he became mute. The mute phase lasted for nine days.
Systematic analysis of orofacial movements was started two days before speech onset (Figure 5.1.). He was cheerful and cooperative. He could drink and eat without problems. He was able to protrude the tongue, but he failed on all other movements. He responded to requests with sighing. He performed on some items of the TROG [23] above age level.
Five days after speech onset, the quality of orofacial movements had improved: he was able to protrude the tongue adequately, and complex movements such as kissing, blowing, and pretending to sense a nasty smell were performed without effort.
Initially speech rate was slightly reduced and articulatory movements were limited. He did not speak spontaneously and gave very short answers. Two weeks later speech was normal.

Case 3. This eight-year-old girl was examined after the resection of a medulloblastoma (Table 5.1.) [1,8]. Postoperatively speech was initially preserved, but two days after the operation consciousness deteriorated. A posterior epidural hematoma was diagnosed and evacuated. After this procedure she remained mute for 60 days.
Assessments were started on postoperative day 50, 10 days before speech onset (Figure 5.1.). The girl lay in bed motionless except for eye movements. She drooled due to the paralysis of the left facial nerve but she could chew and swallow without choking. Occasionally she smiled or produced voiced laughing. She was cooperative and nodded correctly in response to yes/no questions. She could protrude the tongue and make lateral tongue movements but only after considerable latency. She could not initiate other simple or complex orofacial movements. One week later (two days before speech onset) her ability to perform orofacial movements had improved significantly (Figure 5.1.): she could make fast alternating tongue movements, she could close her eyes and show her teeth. The long latency, which had been observed on the previous occasion had disappeared. On requests to make sounds she opened her mouth, but failed to articulate with phonation. As on the first examination, she produced voiced coughs. Two days later, 60 days after the operation, she started to speak. Ten days after speech onset all orofacial movements could be performed adequately.
Initially her speech was moderately dysarthric, characterized by a soft voice, and fluctuating speech rate, pitch, and volume. The dysarthria resolved within one month.

Case 4. (Table 5.1.) In this eight-year-old boy the tumor was almost completely removed with a film of tumor remaining on the floor of the fourth ventricle [1,8]. After surgery he spoke in a normal way, but one day later he became mute. The course was complicated by clinical signs of raised intracranial pressure and pneumonia, for which he was artificially ventilated from day 4 to day 14 postoperatively. After extubation he could swallow properly and eat greedily. He occasionally produced soft whining sounds.

Due to the postoperative complications, the first videotaped assessment took place on postoperative day 35, 26 days before speech onset (Figure 5.1.). He was mute, aloof, and disinterested. He avoided eye contact and had no emotional facial expression. He responded correctly to items of the TROG [23] above age-level. He could be stimulated to write the names of objects. On other requests he fumbled at his bandage in a stereotyped fashion. Ten days later (16 days before speech onset) (Figure 5.1.) he was more compliant and made eye contact. He still had no facial expression. Simple orofacial movements were performed adequately. None of the complex movements could be performed.
On postoperative day 60 his mother reported that he had said 'mama'. One day after the recurrence of speech he was lively and laughed frequently. All simple movements and some complex movements could be performed. Ten days after the recurrence of speech, all movements were performed adequately.
Speech was initially severely dysarthric, characterized by great effort, slow speech rate, high pitch and a remarkable breathiness. Three months later speech was normal except for a slow rate.

Case 5. In this five-year-old boy a large medulloblastoma of the cerebellar vermis was macroscopically completely removed (Table 5.1.) [1]. On the first day after surgery he was extremely apathetic and spoke only a few words, after which he did not speak for five weeks.
We started assessments three days after surgery, 33 days before speech onset (Figure 5.1.). He responded with voiced crying. On a request to repeat the word 'auto' he slightly opened his mouth and then started to cry. Five days later it was hardly possible to examine the boy as he cried almost incessantly. He protruded his tongue successfully once, but after that he refused and turned away abruptly.
Four days later he protruded the tongue and made lateral tongue movements. On other requests for orofacial movements he immediately reacted with coughs resulting in crying. Two days later, 21 days before speech onset, he was able to perform most simple orofacial movements on request. However, he was unable to perform complex movements such as licking his lips, which he had done spontaneously during eating. At this session we tested his language comprehension with the TROG [23] on which test his performance was age-appropriate. Two weeks later (six days before speech onset) we observed considerable improvement: some of the complex movements were now carried out successfully. He was able to make a soundless clicking movement with the tongue. He could make an angry face, but his attempts to chatter with his teeth and lick the lips were not successful. He looked happy and laughed when he successfully performed on tasks.
Speech recurred 36 days after the operation. Three days later almost all orofacial movements were carried out adequately. He was shy when requested to speak. His dysarthria was characterized by a correct but effortful articulation. Words were spelled out at slow rate and with soft voice.

5.5. Results

The scores on the orofacial movement tasks are given in Figure 5.1.. We observed similarities in the performances of the five patients and a similar temporal sequence of recovery. Emotional phonation and chewing were intact throughout the mute phase. Swallowing problems – if at all present – resolved in an early phase of the mute period. Despite adequate function of tongue, lips, oral cavity, and pharynx during eating and drinking, the same movements could not be performed on request. Therefore during the mute phase there is a marked discrepancy between (semi)automatic or involuntary and voluntary orofacial movements, even for simple movements. Complex orofacial movements could rarely be initiated. Children seemed to be aware of this inability as requests for complex movements were usually followed by frustration reaction. Improvement of complex orofacial movements occurred towards the end of the mute phase. Restoration of these complex orofacial movements coincided with the recurrence of speech, as Figure 5.1. shows.

5.6. Discussion

In the present series different mechanisms were taken into account as causes of the mute state. Emotional factors were seriously considered as sick children are reluctant to speak. They may show strong emotional responses to frightening situations as may be the case in operating rooms and intensive care units [25]. At onset of the mute phase, signs of depressed mood such as apathy, lack of spontaneity, negativistic behavior, and frank frustration were indeed frequently present, but during the mute phase, mood improved to such an extent that the children were cooperative or even cheerful on nonverbal tasks. Therefore the speechlessness cannot be attributed to emotional stress, refusal or general unresponsiveness.

Mutism may be the extreme manifestation of aphasia in children [26,27]. A language deficit in our cases is unlikely: Case 1 and Case 4 were able to perform written naming. Language comprehension was adequate in all children during conversation, which impression was confirmed by age – appropriate test performances of Cases 2, 4 and 5 (TROG) [23]. Good language comprehension has also been reported in other studies on cerebellar mutism [2,4,6,7,9].

Dysarthric speech is supposed to mirror the movement disorder of trunk and limbs [24]. However, we could not detect an ataxic component in orofacial movements during the mute phase despite the severe ataxia of trunk and limbs.
Moreover, our analysis of the subsequent dysarthria did not reveal speech features which are characteristic for ataxic dysarthria in adults [24] except for some speech features in Case 3. We conclude that the ataxic motor disorder of trunk and limbs was neither mirrored in orofacial movements during the mute phase nor prominent in speech following the mute phase.

In a recent study [15] an 'apraxic' nature of posterior fossa mutism has been considered. These patients had diffulty 'coordinating oral pharyngeal musculature as manifested by drooling and the inability to swallow' [15]. This so called 'oral pharyngeal apraxia' was present during and beyond the mute phase [15]. We disagree with the term 'oral pharyngeal apraxia' as swallowing cannot be considered a learned movement. Moreover, we did not observe the symptom as reported by Dailey et al. [15]: in all our cases impairment of swallowing – if at all present – resolved in an early stage of the mute phase.

In a broader sense, 'oral apraxia' may be defined as the inability to efficiently and immediately carry out oral movements on command and / or imitation with preserved ability to produce similar actions (semi) automatically [28]. In adults, oral apraxia may be associated with motor speech problems after acquired hemispheric lesions [22]. In these patients the performances are characterized by typical errors such as perseveration and fragmentation of the orofacial movements [22]. This clinical picture is different from that seen in our patients i.e. the loss of the ability to initiate movements without any compensatory crude approximations. It is a matter of definition if our observations fit the criteria for oral apraxia.

We can only hypothesize on the underlying mechanism of our findings. Newer concepts of the role of the cerebellum in motor activity may explain our findings. Ito [29] stresses the cerebellar function in the programming of movement parameters *before* movement initiation. The inititiation of intentional movements may thus be disrupted by cerebellar damage. However, the locus of damage in MSD may not be exclusively cerebellar: in a recent study we demonstrated that multiple factors contribute to the pathogenesis of the MSD syndrome i.e. ventricular location of the tumor, adherence to the dorsal brainstem, preoperative hydrocephalus and edema in the decussation of the superior cerebellar peduncle [1,30]. The SPECT – scan findings in Case 1 suggest a widespread influence of the cerebellar surgical lesion on supratentorial brain structures. The cause of this widespread supratentorial hypoperfusion is not clear. As ventricular size had not increased on MRI in Case 1 in comparison to the preoperative scan, it is hypothesized that the hypoperfusion was caused by diaschisis after the cerebellar trauma. The frontal localisation of this hypoperfusion may explain the 'frontal like' nature of the disorder in our patients i.e. the loss of the ability to initiate orofacial movements in our patients.

For clinical practice we recommend that follow-up throughout the mute phase and after the onset of speech encompasses frequent observation of involuntary and voluntary orofacial movements. The restoration of the latter coincide with the disappearence of the mutism and therefore may carry prognostic value for the time course of the mute phase. Functional imaging as performed in Case 1 may further contribute to an understanding of what Cole [30] designated as the 'foreign policy' of the cerebellum.

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Acquired childhood dysarthria: review on its clinical presentation

6

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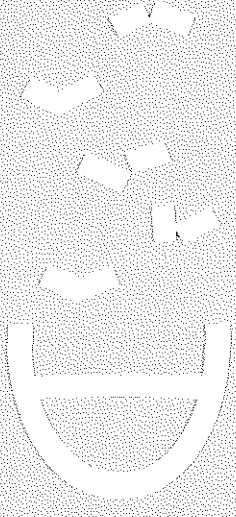

Abstract

The adult classification of dysarthria, correlating with the pathophysiology of the motor systems is usually applied to classify Acquired Childhood Dysarthria (ACD). However, the validity of this adult model for children has not been studied systematically. We reviewed all studies pertaining to analysis of speech features in ACD, published since 1980. Studies were classified on the basis of neuroradiological evidence of lesion site and associated motor disorder. The review demonstrates that knowledge on ACD is based on a limited number of single case studies, of which most pertain to dysarthria following cerebellar tumor resection. Definite similarities with adult dysarthria did not emerge. Some similarity was found for ACD due to basal ganglia lesions. We conclude that ACD needs its own classification.

6.1. Introduction

Dysarthria is a motor speech deficit in neurological disorders. Rate, strength, and coordination of the muscles subserving speech may be impaired to different degrees, affecting articulation, prosody, resonance, respiration, and phonation. Darley et al. [1,2] analyzed speech in 212 adult patients. Deviant speech features occurred in clusters, characterizing major types of dysarthria in adults: ataxic, flaccid, hypokinetic, hyperkinetic, and spastic. The most distinct features of each type as presented by Darley et al. [2] are given in Table 6.1.A.. The authors concluded that these distinctive patterns have localizing value and can assist in identifying the underlying neuropathology, as the deviant speech features mirrored the kind of motor disorder in limbs and trunk [1,2].

Other studies in adults have modified the list of prominent speech features, mainly for ataxic dysarthria [3-6]. Enderby et al. [7] criticized the typology of dysarthria, as pyramidal and corticospinal tracts are modulated by extrapyramidal and cerebellar circuitry prior to excitation of the final common pathways. Consequently, similarities between dysarthrias with different lesions are expected. Despite this criticism, the 'adult model' of distinct dysarthria types [1,2] and their correlation with pathophysiology are widely accepted.

Acquired Childhood Dysarthria (ACD)[1] is certainly not a rare disorder, although an overall incidence or prevalence in pediatric neurology is not known. Despite its frequent occurrence, ACD has hardly been studied systematically (Table 6.1.B.). Most pediatric studies merely report its presence or frequency within the context of a specific neurological condition. Other pediatric studies refer to dysarthria in terms of underlying motor disorder (spastic, ataxic) or neuroanatomic site of damage (cerebellar, bulbar), with no systematic analysis of the speech features. In doing so, these studies implicitly assume the appropriateness of the adult model for children.

The aims of this review therefore are:

1) to survey the literature on speech features in ACD in relation to lesion site and motor disorder, and to discuss the validity of the 'adult model' i.e. the existence of distinct dysarthria types in childhood;
2) to review studies in which the analysis of ACD is of importance for clinical management.

6.2. Inclusion and exclusion criteria

a) We searched Medline databases from 1980 – 1996 for publications available in english under search strategies 'dysarthria' or 'speech disorders'. We selected,

1 For the purpose of the present study we define ACD as a motor speech disorder due to brain lesions acquired in childhood after a normal onset of speech development. We exclude speech disorders in developmental and degenerative disorders.

Table 6.1. Comparison of features in adult dysarthria types with pediatric cases.

Table A. Most distinctive speech features in adult dysarthria types [2]	Table B. Studies reporting features in children
ATAXIC DYSARTHRIA	
– irregular breakdown of articulation	[17]
– excess and equal stress	[14,23]
– prolongations of phonemes and intervals	[-]
– slow rate	[13,15,17-19,21,22]
– harsh voice	[-]
– monotony	[12, 13, 15, 19]
FLACCID DYSARTHRIA	
– hypernasality	[25]
– nasal emission of air	[-]
– continuous breathiness	[25]
– audible inspiration	[-]
HYPOKINETIC DYSARTHRIA	
– monopitch	[-]
– monoloudness	[-]
– hypophonia	[-]
– imprecise articulation	[33,35,36]
– variable rate	[33,35]
– short bursts of speech (stuttering)	[33,35]
QUICK HYPERKINETIC DYSARTHRIA (CHOREA)	
– hypernasality	[-]
– harshness	[-]
– breathiness	[-]
– loudness variations	[-]
SLOW HYPERKINETIC DYSARTHRIA (DYSTONIA)	
– imprecise articulation	[34]
– hypophonia	[34,35]
– variable rate	[-]
– harshness + strained strangled voice	[-]
– transient breathiness + audible inspiration	[-]
SPASTIC DYSARTHRIA	
– imprecise articulation	[-]
– slow rate	[-]
– low pitch	[-]
– harsh voice	[-]
– strained strangled voice	[-]
– pitch breaks	[-]
– reduced stress	[-]

[..] Reference number

[-] Not reported in children

reviewed and analyzed studies pertaining to acquired dysarthria;

b) We included only descriptions of children ≤ 16 years;

c) We included studies mentioning perceptual judgment of speech features according to the Mayo Clinic List [1,2] shortly after the onset of the speech problem. These studies are summarized in Tables 6.2A., 6.2B., 6.3., and 6.4.. To illustrate a clinical context in which ACD may occur, we sometimes refer to cases with nonspecific indications such as 'slurred' or 'dragged' speech, but these cases are not included in the Tables;

d) Cases were excluded when the speech disorder was a mixture of dysarthria and aphasia or verbal apraxia, as this obscures the very nature of the dysarthria[1].
We classified on the basis of neuroradiological evidence of lesion site and associated motor disorder, thereby classifying according to the motor subsystems.

6.3. ACD in relation to site of damage and motor impairment

6.3.1. Cerebellar lesions

Speech features of cerebellar dysarthria in adults are given in Table 6.1.A.. In children, cerebellar tumor resection may cause a syndrome of mutism with subsequent dysarthria, further referred to as MSD. This clinical picture has been described in a greater number of children than ACD associated with any other lesion site. The primary site of lesion is the cerebellar vermis but the tumor and the effects of surgery may affect adjacent structures. Most studies refer to type and severity of dysarthria in the acute stage after recovery from mutism. In 20 cases, speech was judged according to the Mayo Clinic List of speech features by experienced professionals or speech pathologists. These studies are mentioned in Table 6.2A..
Table 6.2A. shows that all children with MSD [11-23] had severe limb and trunk ataxia. *Slow articulation, monotony* and *hoarse soft voice* are the most frequent speech abnormalities. Irregular articulatory breakdown and excess and equal stress resulting in scanning speech – most prominent in ataxic speech in adults (Table 6.1.A.) – , were certainly not the hallmark of ACD in these children. Nagatani et al. [15] and Al Jarallah et al. [19] explicitly mentioned that speech was *not explosive nor scanning.* Van Dongen et al. [18] found that the observed speech features did not fit into any specific cluster of deviant speech dimensions. In the course of improvement there was a marked dissociation between speech impairment and motor disability: the initially severe dysarthria recovered (almost) completely while ataxic motor impairment persisted in most children.

We could find only two reports on speech characteristics of ACD following cerebellar damage with a different etiology (Table6.2B.).

Echenne et al. [24] reported swallowing difficulties, velar paralysis, and *dysphonic, hoarse speech* of short duration after two episodes of vomiting, headache, and

1 For differential diagnostic issues we refer to other studies [8-10].

Table 6.2A. Clinical data of children with mutism and subsequent dysarthria (msd) after cerebellar tumor resection.

Study	Sex/age	Lesion site	Motor disorder	Lower cranial nerves	Speech features
Rekate [11]	F/8	medial	limb ataxia, R paresis	N	slow, monotonous.
Volcan [12]	F/8	medial	trunk ataxia, R paresis	L VI paresis	monotonous, monosyllabic.
Dietze [13]	M/7	RH and LH	trunk ataxia	N	sparse, slow, monotonous.
Ferrante [14]	F/5	medial	ataxic gait	N	scanning.
Nagatani [15]	F/4	medial	trunk ataxia	N	slow, monotonous, not explosive nor scanning.
Catsman [16]	M/6	medial	trunk and limb ataxia, R paresis	L/R XII paresis	audible respiration, incoordination of respiration and speech, hoarse, nasal, consonant distorsion.
Herb [17]	M/9	medial	R ataxia R paresis	L/R VII paresis	aphonia, slow articulation, no synchronisation of articulation and phonation.
Van Dongen [18]	F/8	medial	trunk and limb ataxia	L VII paresis, L XII paresis	alternating loudness, flat vowels, consonant cluster reduction.
Van Dongen [18]	M/8	medial	trunk and limb ataxia, R paresis	L XII paresis	hoarse, soft voice, omission of final sounds.
Van Dongen [18]	M/5	medial	trunk and limb ataxia	XII weakness	slow rate, strained-strangled voice, syllabification.

Al Jarallah [19]	F/9	medial	N	N	slow, monotonous, *not* explosive.
Crutchfield [20]	M/7	medial	upper limb and trunk ataxia	N	whispering.
Kingma [21]	M/5	medial	L hemiparesis	L VII paresis	slow, weak, monosyllabic.
Kingma [21]	M/9	4th Ventricle	R ataxia	?	slow speech rate.
Pollack [22]	M/16	medial	L paresis	N	high pitched nasal voice.
Pollack [22]	M/9	medial + 4th Ventricle	L ataxia	L VI paresis	high-pitched voice.
Pollack[22]	F/6	medial + 4th Ventricle	R ataxia	N	whispering, monosyllabic, slow.
Asamoto[23]	F/8	medial	dysmetria	N	monotonous, monosyllabic, scanning.

Table 6.2B. Clinical data of children with cerebellar lesions.

Study	**Sex/age**	**Etiology**	**Motor disorder**	**Lower cranial nerves**	**Speech features**
Echenne [24]	M/9	infarction L cerebellar H	L hemichorea and ataxia, R paresis	IX/X paresis	dysphonia, hoarseness
Dietze [13]	F/15	resection of arterio-venous malforma-tion R and L cerebellar H	diffuse hypotonia, bilateral and truncal ataxia	N	monotonous, labored and bradykinetic

F = female; M = male; R = right; L = left; H = hemisphere; N = normal; ? not reported

dizziness (Table 6.2B.). CT scan showed an area of decreased density in the lateral part of the left cerebellar hemisphere, caused by a filling defect of the distal portion of the basilar artery. Dietze and Mickle [13] observed ACD after the resection of a cerebellar arteriovenous malformation, but the features did not resemble those of adult dysarthria after cerebellar lesions (Tables 6.1. and 6.2B.).
Conclusion: Dysarthria, following a mute phase after cerebellar tumor resection is frequently characterized by slow speech rate, monotony and hoarse soft voice. Table 6.1. shows that excess and equal stress, distinctive for ataxic dysarthria in adults, was only found in two cases. In MSD, dysarthria usually disappears, whereas ataxia persists in most children for a considerable time. The cited studies do not sufficiently support current practice to label dysarthria in MSD ataxic. In addition, cerebellar lesions of a different etiology equally did not cause speech features resembling ataxic dysarthria in adults.

6.3.2. Brainstem lesions

Damage to the nuclei of the lower cranial nerves in the bulbar region of the brainstem may result in flaccid bulbar dysarthria. Prominent speech features of adult bulbar dysarthria [1,2] are given in Table 6.1.A.. For the purpose of the present study, we excluded diseases affecting the (peripheral) neuromuscular junction or the muscles as is the case in myasthenia gravis, muscular dystrophy or Guillain Barré Syndrome. We reviewed studies with neuroradiological evidence of brainstem lesions associated with ACD (Table 6.3.).

Strokes due to vertebral artery occlusion, a well-known entity in adults are rare in the pediatric population [27,28]. Older studies reported *slow, slurred,* or *dragged* speech [27,29]. One study reports on speech features after occlusion of the basilar artery in a six-year-old boy [25]. ACD in this child was associated with bulbar palsy. The speech pattern resembled flaccid dysarthria in adults. Initially, CT-scan showed no abnormalities, but some weeks later, a small infarct in the left thalamus was demonstrated.

The clinical picture of mutism and subsequent dysarthria, frequently labeled cerebellar, has also been reported after the surgical resection of a cavernous hemangioma of the right pons at the level of the middle cerebellar peduncle [26] (Table 6.3.). *Impairment of prosody and rhythm of speech* was labeled 'cerebellar' dysarthria.

Two children of a series of children with posterior fossa tumor did not manifest mutism but did have mild speech problems after surgery [18] (Table 6.3.). A 10-year-old boy spoke with a very *soft voice* two days after extubation following resection of a tumor in the fourth ventricle, invading the right cerebellar hemisphere and the right cerebellar peduncle [18]. A four-year-old girl answered questions by silently moving her lips and *whispered* a limited number of words, following resection of a meningioma, situated anterior to the basilar artery [18].

Table 6.3. Clinical data of children with brainstem lesions.

Study	Sex/age	Etiology	Motor disorder	Lower cranial nerves	Speech features
Bak [25]	M/6	infarct left posterior thalamus	L paresis	L VII, L IX/X, L XII pareses	imprecise consonants, distorted vowels, hypernasality, breathy voice, harsh voice, monopitch, monoloudness
Van Dongen [18]	M/10	IV th ventricle tumor	mild limb ataxia	R IX,X, and XII paresis	very soft voice
Van Dongen [18]	F/4	posterior fossa tumor anterior to the basilar artery	flaccid tetraparalysis	N	whispering a limited number of words
Frim [26]	F/8	surgical resection of pontine cavernous hemangioma	L paresis	L VII weakness	dysarthria of speech prosody and rhythm

F = female; M = male; R = right; L = left; N = normal; ? = not reported

In adults, speech deficits following bilateral thalamic lesions have been documented by Ackermann et al. [30]. In children, little is known about speech deficits after thalamic lesions. Garg and DeMyer [31] presented the clinical features of six children with thalamic strokes, of whom one child had *slurred speech.* Parker et al. [32] also reported *slurred speech* after infarction in the left thalamus associated with mycoplasma infection in an eight-year-old girl, but no further details on the speech features are given.
Conclusion: One Case study [25] revealed a cluster of deviant speech features after brainstem lesion with clinical signs of bulbar palsy, resembling flaccid bulbar dysarthria in adults. Impairment of prosody was reported after resection of a cavernous hemangioma of the pons [26].

6.3.3. Basal ganglia lesions

In adults, movement disorders associated with basal ganglia lesions fall into two categories with opposite symptomatology: hypokinesia and muscle rigidity respectively hyperkinesia with choreatic or dystonic movements. These two clinical entities are well recognized in speech as hypokinetic and hyperkinetic dysarthria [1,2], as is shown in Table 6.1A..

In children, pure hypokinetic or hyperkinetic movement disorders seldom occur. More often, there is a mixed extrapyramidal syndrome.
Characteristics of children with basal ganglia lesions and associated speech features are given in Table 6.4..
Murdoch et al. [33] presented a 13-year-old boy with post-hypoxic hypokinetic movement disorder. Detailed speech analysis during a long follow-up period after a mute phase revealed the following speech features: *difficulty in controlling speech rate within a phrase,* and *increase of speech rate within a sentence* (Table 6.4.). These features were labeled hypokinetic dysarthria, as they are highly specific for adult Parkinson patients and do not occur in any other dysarthria type.
We found one case of ACD associated with hyperkinetic dystonic and choreiform movement disorder [34](Table 6.4.). Al-Mateen et al. [34] described a nine-year-old girl with encephalitis due to mycoplasma infection. MRI showed areas of low-signal intensity involving the caudate, the putamen, and the globus pallidus. One month after disease onset, she had dystonic posturing of the right foot, dyskinetic movements of the face, and choreiform movements of the extremities. Speech was *hypophonic with incomprehensible vocal utterances.* In the course of improvement, a marked discrepancy between severity of speech deficit and movement disorder was noted: 18 months after onset of the illness, 'she was still unable to initiate speech without cuing but she could run, skip and climb' [34].
Mixed hyper- and hypokinetic movement disorder associated with ACD has been described in detail in 2 cases [35](Table 6.4.). Pranzatelli et al. [35] examined six cases with extrapyramidal movement disorders due to various etiologies. Abnormal speech was present in all children, of whom three were initially mute. Case 1, a 12-year-old girl, virtually unable to move or speak, improved with medication and produced

Table 6.4. Characteristics of children with basal ganglia lesions.

Study	Sex/ age	Etiology	Motor disorder	Lower cranial nerves	Speech features
Murdoch [33]	M/13	postanoxic bilateral striato-capsular lesions	clinical features of parkinsonism	L/R VII paresis	7 weeks:impairment of bilabial consonants, slow rate, labored speech; 10 weeks: difficulty controlling speech rate, impaired initiation of speech
Al-Mateen [34]	F/9	inflammatory lesions of caudate nucleus, putamen, and globus pallidus	dystonia, choreiform movements of the extremities	?	hypophonic incomprehensible utterances
Pranzatelli [35]	F/12	anaphylaxic shock	dystonia, bradykinesia, rigidity	?	after mutism hypophonic speech
Pranzatelli [35]	M/14	head trauma	dystonia, hemiballismus, bradykinesia, rigidity	?	stuttering speech
Aram [36]	F/8	vascular lesion globus pallidus, putamen, posterior part of internal capsule, caudate	R paresis	N	consonant cluster reduction, consonant omissions

F = female; M = male; R = right; L = left; N = normal; ? = not reported

hypophonic speech (Table 6.4.). Case 2, a 14-year-old boy, suffered from bradykinesia and rigidity, but in addition he had writhing hemiballistic movements of the left hand. This mixed extrapyramidal movement disorder was associated with *stuttering speech* (Table 6.4.).

In one case, basal ganglia lesions were associated with ACD but not with extrapyramidal movement disorder. Case 2 of Aram et al. [36] (Table 6.4.) exhibited dysarthria following a left-sided infarction involving the globus pallidus, the putamen, the body of the caudate, and the posterior limb of the internal capsule. She had a right-sided hemiparesis, but no signs of extrapyramidal movement disorder. Speech was mildly affected, mainly in articulation. Silverstein and Brunberg [37] reported a mild dysarthria with rapid resolution in two children with postvaricella basal ganglia infarction. One child presented with hemiparesis and hemichorea, associated with *slurred speech.*

Conclusion: The review shows some resemblance with the adult model. In children with extrapyramidal movement disorders, dysarthria may be characterized by hypophonia, stuttering, and difficulty in controlling speech rate which are highly specific for hypokinetic dysarthria in adults (Table 6.1.). Moreover, the degree of movement disorder appears to correlate with the speech impairment except for one case [34]. Both may subside on medication, which finding substantiates the correlation of the extrapyramidal movement disorder with the severity of the dysarthria.

6.3.4. Cerebral cortical lesions

Anterior opercular syndrome

Functional impairment or structural damage to the anterior part of the insula, the operculum, leads to the well-studied opercular syndrome with uniform symptomatology. Its most significant hallmark is the loss of voluntary control of orofacial musculature with drooling, anarthria (i.e. speechlessness as most severe form of dysarthria) and aphonia. Mental status is preserved and language comprehension is intact. There is no emotional lability. Voluntary orofacial movements are impossible on request, whereas patients may yawn, laugh, or be able to perform complex movements spontaneously, such as blowing out a match [38]. In adults, the opercular syndrome is most often attributable to cerebral ischemia. The major deficits – dysphagia and anarthria – are unlikely to be reversible, as no full recovery is on record [38].

In children, various etiologies may cause the (rare) opercular syndrome. Because of its uniform clinical picture, data are not summarized in a table. Deonna et al. [39] reported on oromotor and speech disturbances as a focal epileptic manifestation. They examined three children, diagnosed with Benign Partial Epilepsy with Rolandic Spikes. Due to recurring seizures around the sylvian area, *speech arrest* of short duration, *slow speech rate, poor prosody* and *imprecise articulation* occurred as prolonged or postictal deficits, sometimes resembling an incomplete anterior opercular syndrome. Shafrir and Prensky [40] observed opercular syndrome of

epileptic origin in a child with recurrent prolonged episodes of severe oral apraxia, dysarthria and drooling. During these episodes there was an increase of right temporal spikes and appearance of generalized spikes, polyspikes and slow wave discharges. Remissions were associated with improvement in the EEG, which either showed right temporal focus or was free of epileptiform activity [40].
Continuous partial seizures in the rolandic areas during a prolonged period of time caused extreme drooling, impairment of tongue function, and *slurred speech* in two children with preexistent Benign Partial Epilepsy with Rolandic Spikes (BPERS). Speech recovered fully after adequate treatment of the seizures [41].
Other etiologies as a cause of opercular syndrome have been described in children. A 13-year-old boy [42] sustained biopercular infarctions following vasculitis after a scorpion bite. The mutism lasted for months, after which he was able to laugh and cough and he could produce a few isolated vowels. Massive infarctions in both opercula after tuberculous meningitis also caused opercular syndrome in a three-year-old boy, who became verbally mute and aphonic, with his mouth constantly open and dribbling saliva [43]. One year later he had regained the ability to articulate simple words. The patient of Prats et al. [44] who manifested opercular syndrome after herpes simplex encephalitis, remained completely mute. Recovery from opercular syndrome after infectious disease was also poor in two other children [45]. At follow-up after two years, Case 1 was able to speak with *impaired articulation* and *nasal speech.* At onset, Case 2, a 24-months old girl, had complete absence of speech and of all voluntary movements of jaw, lips and tongue. Two years later, she had not regained any speech except for sounds and *a few and poorly articulated words* [45].
Conclusion: On an ictal or postictal basis, functional impairment of the anterior opercula interferes with orofacial musculature and speech. The mode of onset, relapses and evolution show various patterns [39]. Outcome is favourable, once the epilepsy is adequately treated.
After structural lesions the clinical picture is similar to that seen in adults. The prognosis is poor, as is the case in adults: patients remain mute for a long period of time and rarely regain speech. Speech features have therefore been incompletely analyzed.

Other cerebral cortical and internal capsule lesions
Deviant speech features in adults with spastic dysarthria, occurring after cerebral cortical and internal capsule lesions, are presented in Table 6.1.A.. Bilateral damage may result in persistent speech impairment in adults. Accompanying neurological deficits consist in a pyramidal syndrome on one or both sides.

In children, dysarthria without aphasia or verbal apraxia [8-10] after bilateral cerebral hemisphere lesions, that are not located in the opercula, have rarely been documented. Data are not summarized in a Table, as we could find only one case, which fits our inclusion criteria [46]. *Stuttering-like speech* was noted in this 27-months-old child: she suffered from multiple infarctions in the periventricular white matter, 14 months after she had sustained a subcortical infarction in the left basal ganglia that extended into the cortex [46]. Her stutter, which lasted for seven weeks, affected mainly the initial phonemes and speech was *hypophonic.*

Unilateral hemisphere lesions, on the other hand, have been reported to cause ACD, but descriptions of speech features were less specific. Following varicella infection, a nine-year-old boy sustained a left cerebral infarction involving the internal capsule and the caudate nucleus [47]. He became dysarthric, and developed a right-sided central facial palsy and a right-sided hemiplegia, affecting the upper limb. All neurological features resolved completely within hours [47].
Slurred speech and hemiparesis of the right arm and leg were the symptoms in a six-year-old boy, who sustained a small left middle cerebral artery ischemic infarction extending into the subcortical white matter [48]. His clinical status showed fast improvement with complete recovery after three months [48].
Conclusion: Descriptions of ACD following circumscribed cerebral hemisphere lesions are scarce. As unilateral lesions may lead to ACD of transient nature, studies have not focused on its specific features. Bilateral non-opercular hemisphere lesions associated with dysarthria, frequent in adults, were only documented in one child, who developed stuttering-like speech impairment [46]. To our knowledge, dysarthria without aphasia or verbal apraxia, following acquired non-opercular cortical lesions and resembling spastic, pseudobulbar dysarthria in adults has not been described in children.

6.4. The presence of ACD in clinical management

In the previous paragraphs, we studied the relationship between ACD on the one hand, and motor disorder on the other, according to the clinical symptomatology or the neuroradiological evidence of lesion site.
The second aim of this study was to review studies which demonstrate that the occurrence of dysarthria may be a marker for clinical deterioration or that recovery from dysarthria may herald clinical improvement. The analysis of dysarthria may therefore be of importance for clinical management.
Reitman et al. [49] analyzed speech deficits in survivors of Reye's syndrome. Twenty-six of the 43 examined children were dysarthric, aphonia being most frequently noted. In four children the dysarthria was the predominant handicap at 12 months follow-up. The authors concluded that rehabilitation of Reye's syndrome should anticipate the occurrence of speech problems.
Intrathecal Methotrexate (IT MTX) treatment is an essential part of CNS prophylaxis for leukemia in children. Neurological complications due to neurotoxic reactions may follow such therapy [50,51]. Yim et al. [51] observed severe hemiparesis and brief but profound dysarthria of sudden onset in two children after IT MTX treatment. Initially, these symptoms were considered part of a MTX-induced encephalopathy. Later on, CT and MRI showed ischemic structural lesions in the cerebral hemisphere in both children. This clinical picture, in which the profound dysarthria was prominent, may be an indication for ischemic brain lesions, and differs from the clinical picture of MTX encephalopathy.
The presence of dysarthria seems to have a prognostic value in cases of thalamotomy, performed to alleviate the disabling tremor which may occur in posttraumatic midbrain syndrome. In two patient groups (aged 10-29 years) [52,53], thalamotomy

improved the tremor. However, the preexistent posttraumatic dysarthria tended to worsen postoperatively. Both studies [52,53] mentioned the worsening of the dysarthria as a major limitation to the surgical intervention and concluded that the existence of a preoperative dysarthria is a contraindication for thalamotomy.
Van Dongen et al. [54] analyzed dysarthric speech features in children with supranuclear or peripheral facial palsy. (As lesions were diffuse, we did not include this study in the previous paragraphs.) Within the present context it is worth mentioning as severity of dysarthria was analyzed in relation to neurological deficits. In patients with supranuclear palsy, recovery from severe dysarthria to intelligible speech preceded improvement of other motor functions.
Conclusion: Dysarthria may occur as an initial symptom of rare structural lesions during MTX treatment. It may signal structural brain damage as a result of ischemia. As such, its sudden onset as a focal neurological feature differs from the neurotoxic reactions in MTX encephalopathy. The recognition of the sudden onset of dysarthric speech, is therefore, of diagnostic significance, and contributes to clinical management [51]. In thalamotomy performed to alleviate posttraumatic tremor, the dysarthria may worsen despite improvement of other motor function. The presence of dysarthria constitutes a major contraindication for thalamotomy [52,53]. Recovery from dysarthria may precede neurological improvement [54].

6.5. Discussion

The main aim of this study was to provide a survey of speech features in ACD and to compare these with the adult types of dysarthria [1,2]. In children, dysarthria associated with cerebellar tumor resection [see Table 6.2A.] has received more attention than dysarthria associated with any other type of lesion. Within this group, speech was frequently characterized by slow speech rate and monotony. These features are not distinctive for ataxic dysarthria as they may occur in other dysarthria types. Excess and equal stress – scanning speech –, considered the hallmark of ataxic dysarthria in adults and not occurring in any other dysarthria type [1,2], was rarely observed, even in children with severe dysarthria. We found one other case of ACD after cerebellar stroke [24], which did not show typical ataxic speech features either. Thus, in children, cerebellar lesions, particularly tumor resection, may cause speech deficits, not resembling ataxic dysarthria in adults. As yet, the validity of the adult model for children with cerebellar lesions has not been sufficiently proven.

In other patient groups with lesions in different areas of the brain, grossly classified on the basis of neuroradiological or neurophysiological similarities, clusters of deviant speech features that would be specific for each group have not emerged either. An exception appears to be the speech features associated with basal ganglia lesions [33-37]. Features that are not reported in other dysarthria types, such as short rushes of speech, difficulty in controlling speech rate (festination) and stuttering were also recorded in children (Table 6.1.). This suggests that lesions involving the basal ganglia may cause speech deviancies that resemble dysarthria in adults with similar movement disorders.

We had two reasons to take the 'adult model' of clusters of speech features mirroring the motor impairment, as a starting point [1,2]. Firstly, these studies comprise the largest groups of dysarthric adults ever investigated. The typology of dysarthrias [1,2] has been applied universally to label deviant speech, in adults as well as in children. Secondly, an equivalent model of motor speech disorders for children is lacking. Therefore, Murdoch et al.[55] claim that although it is difficult to apply models and theories developed for adults to children, it appears to be appropriate to use the classification system of Darley et al. [1] to describe the equivalent speech disorders in children, until more information becomes available to refute its appropriateness.
In the light of this, it is of importance to stress that the quality of nonspeech movements, articulation, velopharyngeal function, and orofacial musculature change during development [56,57]. Moreover, normal speech in children may reveal features such as strained voice, breathiness and hyponasality that are considered pathological in adult speech [58]. Developmental norms are well known to speech pathologists working with developmentally speech/language impaired children, but may be poorly recognized within clinical circles. We could find only one study addressing the discrimination of developmental and acquired dysarthric aspects of articulation in acquired cerebellar lesions [59], but this study did not fit our inclusion criteria.

As to the second aim of our review, the available literature shows that analysis of dysarthria may play a crucial role in the recognition of clinical changes.

All observations, reviewed in the present study, are based on single case studies. This illustrates the fact, that dysarthria has been treated as a Cinderella [60], in children even more so than in adults. As yet, the study of ACD is still in its infancy, as was the case with the study of acquired childhood aphasia in the late 1970s. Until then, studies on aphasia in children had focused mainly on the fundamental differences between the neurology of language in children and adults [61]. More recently, systematic studies of recovery from acquired childhood aphasia and of lesion analyses (reviewed by Paquier and Van Dongen [8]) show that correlations between aphasia type and lesion localization parallel those found in adults [8,61]. As to ACD, it is not yet possible to predict what systematic analyses of speech features and anatomo-clinical correlation studies will teach us about the similarity and differences between dysarthria in children and in adults. The current knowledge on ACD, reviewed here, shows that ACD needs its own classification through detailed analysis of speech features in dysarthric children.

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Dysarthria in children with cerebellar or brainstem tumors: does it sound differently?

7

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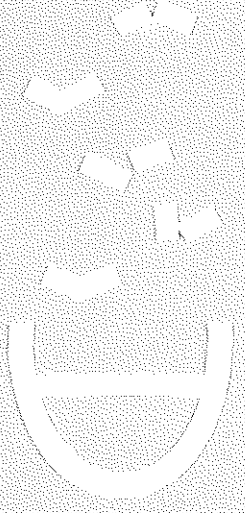

Abstract
We perceptually analyzed speech features in two groups of children. The first (N=6) had undergone cerebellar tumor resection, the second (N=6) comprised children with a brainstem tumor. Children belonging to the first group became dysarthric after a mute phase. Slow speech rate was a specific feature, but scanning speech and irregular articulatory breakdown – i.e. prominent characteristics in adult ataxic dysarthria – were not observed.
In the second group, hypernasality was a prominent characteristic; this resembled flaccid dysarthria in adults. These findings suggest that acquired childhood dysarthria needs a proper classification.

7.1. Introduction

Dysarthria is a motor speech deficit in neurological disorders. Rate, strength, and coordination of the muscles subserving speech may be impaired to different degrees affecting pitch, loudness, vocal quality, respiration, prosody, and articulation. Darley et al. [1] presented a list of 38 items to judge speech, now universally accepted [2]. Darley et al. [1] perceptually analyzed these 38 speech features in 212 dysarthric adults. They found that items occurred in clusters, constituting different types of dysarthria: flaccid dysarthria (in bulbar palsy), spastic dysarthria (in pseudobulbar palsy), ataxic dysarthria (in cerebellar disorders), hypokinetic dysarthria (in parkinsonism), and hyperkinetic dysarthria (in dystonia and chorea). Other authors subsequently modified the cluster model of Darley et al. [1], especially regarding ataxic dysarthria [3,4].

In children, little is known about the characteristics of dysarthric speech in various acquired neurological disorders despite the fact that Acquired Childhood Dysarthria (ACD) frequently occurs. A recent review [5] shows that most pediatric studies merely report the presence of ACD within the context of a specific neurological condition, and that but a limited number of pediatric studies, mainly pertaining to dysarthria following cerebellar tumor resection [5], analyzed speech in accordance with Darley et al. 's [1] method. To our knowledge there is only one study documenting dysarthric speech features in a child with brainstem lesion [6]. Thus, there is a lack of studies describing speech features in children with ACD.

The purpose of this study, therefore, is to identify speech features in two groups of children with similar etiology (tumoral) but with distinct site of lesion (cerebellar vs. brainstem).

7.2. Patients

We examined 12 children, six with cerebellar and six with brainstem tumors. In all children, the site of the tumor was documented by MRI during hospital stay.

Cerebellar tumor group

In the period 1989-1996, 44 children were admitted for cerebellar tumor resection to the University Hospital Rotterdam. Twelve of these children postoperatively suffered from the syndrome of 'mutism and subsequent dysarthria', of which group we excluded six patients from this study: three were too young for proper speech evaluation (three-years-old), one was too old for inclusion in the pediatric group (17-years-old), one was a nonnative speaker, and in one child dysarthria resolved too rapidly to be recorded properly. Table 7.1. displays the clinical data of the remaining six patients. All children had had a normal psychomotor development, and preoperatively none of them was dysarthric. All children were examined in the acute phase of the dysarthria, two to four weeks after the disappearance of the mutism.

Table 7.1. Clinical data of two groups of children with tumors.

Case	Sex/age	Tumor type	Cerebellar			Pyramidal		Cranial nerves	
			oculo	trunk	limb	left	right	left	right
A. Children with cerebellar tumors									
A1	M/8	medulloblastoma	+	++	++	N	+	N	N
A2	F/8	medulloblastoma	+	++	++	++	++	VII	N
A3	M/6	medulloblastoma	++	++	++	N	++	VI	VI
A4	M/5	medulloblastoma	N	++	++	N	++	N	N
A5	M/12	medulloblastoma	++	++	++	++	+	VI,IX	IX
A6	M/12	medulloblastoma	++	++	++	++	N	II	II
B. Children with brainstem tumors									
B1	M/14	glioblastoma	++	N	++	++	N	VI,VII	VI
B2	M/11	astrocytoma	N	N	N	+	N	V	VI
B3	F/11	atrocytoma	++	N	+	++	N	N	VI,VII,VII
B4	F/12	glioma	+	N	++	+	N	VII	N
B5	M/4	glioma	N	N	+	++	+	III,VI	III,VI,IX
B6	M/7	astrocytoma	++	N	+	+	+	IX	IX

M=male;F=female;N=normal;+=mildly abnormal;++=severely abnormal.

Brainstem tumor group

In the same period, speech of six children with brainstem tumors was analyzed. Table 7.1. displays their clinical characteristics. These children also had had a normal psychomotor development before disease onset. Dysarthria was present as an initial symptom at time of diagnosis, one to 16 months prior to speech analysis. Cases B1, B3, B5, and B6 were treated with irradiation, and Case B3 also with chemotherapy.

7.3. Methods

Speech was perceptually analyzed on 36 of the 38 speech features of Darley et al. [1]. We excluded global ratings for intelligibility and bizarreness. Three authors (MVM, EYB, HRVD) independently rated speech features from audiotaped or videotaped samples of spontaneous speech and repetition.

We used the University of Michigan rating system [4], which ranges from 0 (unaffected) to 3 (severely affected). In case of disagreement, samples were relistened to until consensus was reached. For further details on the method we refer to earlier studies of our research group [6-8].

7.4. Results

A number of speech features[1] were not met in any of the children. This was not surprising for some of them, as they only occur in connection with specific diseases not included here. For instance, short rushes of speech are mainly found in Parkinson's disease [1].

Table 7.2. shows the results on the remaining speech characteristics. We discuss the most conspicuous results. In both groups, imprecise consonants and distorted vowels were perceived to various degrees. However, within the context of these articulatory impairments we also found striking differences.

In the cerebellar group, slow speech rate was prominent and distinctive (Table 7.2.), as analysis revealed a significant group difference ($p < .05$, Fisher exact probability test). Other speech features, thought to be characteristic of ataxic dysarthria [1], were only mildly impaired, if at all present.

In the brainstem group, in addition to the previously mentioned articulatory impairments, hypernasality appeared to be a conspicuous speech feature, being more frequent and more severe than in children with cerebellar lesions. Analysis of frequency alone did not reveal significant group difference. Respiration was severely impaired in only one child (Case B3).

1 Voice tremor, excess loudness variation, voice stoppages, hyonasality, nasal emission, grunt at end of expiration, increase of rate in segments, increase of overall rate, reduced stress, prolonged intervals, short rushes of speech, excess and equal stress, repeated phonemes, irregular articulatory breakdown.

Table 7.2. Speech features[1]

Speech feature	Cerebellar tumor group						Brainstem tumor group					
	A1	A2	A3	A4	A5	A6	B1	B2	B3	B4	B5	B6
Pitch												
1a. low pitch			*				*					
1b. high pitch	*	*		*								*
2. pitch breaks									*			
3. monopitch	**		*									
Loudness												
5. monoloudness	*											
7. loudness decay							*		**	*		
8. alternating loudness		**										
9a. soft voice								*	**			**
Vocal quality												
10. harsh voice	**				*		*					*
11. hoarse wet voice	*										**	*
12. breathiness									**	*		
14. strained voice		*										
16. hypernasality					*	*			***	**	***	*
Respiration												
19. forced in-expiration									**	*		
20. audible inspiration			*						***			*
Prosody												
22a. slow rate	**	*	***	**	**	**					***	
23. short phrases									**			*
27. variable rate						*						
29. inappr. silences							*		**			
Articulation												
32. imprecise consonants	*	*	**	*	*	**	*	*	**	**	***	*
33. prolonged phonemes						**						
36. distorted vowels		*	**		*	**		*		*	***	*

1 The numbering of speech features corresponds with list of speech items of Darley et al. [1]* = slightly impaired; ** = moderately impaired; *** = severely impaired.

7.5. Discussion

We analyzed speech characteristics in children with cerebellar or brainstem tumors. The set of data is limited, because sample sizes were small but etiology was the same in all children. Ataxia, pyramidal signs and cranial nerve dysfunction were present to various degrees in both groups, which is in agreement with the clinical descriptions of these patient groups [8,9].
We found articulatory impairments in both groups, but also specific differences: the dysarthria sounded differently in the two groups, because of slow rate in the cerebellar and hypernasality in the brainstem group. These findings are in agreement with other pediatric studies. Studies on dysarthria following cerebellar tumor resection also mention slow rate [5]. We could not find reports on ACD due to brainstem tumors. One study [6] reports speech deficits in a six-year-old boy with a lesion to the left posterior thalamus of vascular etiology. His speech impairment resembled our observations in the brainstem group.

In the light of the scarcity of reports on ACD, we wondered if our results are in accordance with those found in adults. Considering the role of etiology in speech disorders, we wanted to compare our findings with data collected in adult patient groups with similar etiology. Unfortunately, we could not find such studies. Concerning the cerebellar group, we could only compare our data with those recorded in adults with various etiologies [1,3,4]. As was the case in our cerebellar group, Ackermann et al. [3] also found that slow speech rate was the most common dysarthric speech feature after ischemic cerebellar lesions. According to other studies [1,4], excess and equal stress -scanning speech- and irregular articulatory breakdown are characteristic of adult ataxic dysarthria. The absence of such speech features in our cerebellar group is a remarkable finding. Ackermann et al. 's [3] suggestion that scanning speech might occur in more severe types of cerebellar dysarthria was not confirmed by our observations in children: scanning speech was neither observed during the early phase of severe dysarthria, nor later on.
Our data partly confirm the adult model of flaccid dysarthria by the presence of hypernasality and imprecise consonants in the brainstem group. Other speech features, prominent in adult flaccid dysarthria [1] were rarely heard: continuous breathiness was only perceived in two cases (B3 and B4) to a mild or moderate degree; monopitch did not occur in any of the children with brainstem tumors.

The differences between adults and children with cerebellar or brainstem lesions may be explained by age or etiology. Lesions incurred during maturation of speech motor control may result in speech deficits, different from adults'. We are reluctant to generalize our findings. The tumoral etiology does not enable to clarify which (sub)structures contribute to the observed speech disorders. Speech features in children with well circumscribed vascular lesions may elucidate this point, but such lesions constitute uncommon events in childhood.

We conclude that Darley et al. 's [1] list of speech items is a good instrument to describe ACD. However, the adult cluster model [1] is not suitable for the pediatric

age group. An age-related classification system should be developed. This study is a first attempt to classify ACD – thus far treated as a Cinderella by language pathologists [10] –, as it evidenced speech characteristics, associated with distinct lesions of identical etiology in two pediatric groups.

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8

General discussion

8.1. Introduction

Since its first description [1] the syndrome of mutism and subsequent dysarthria (MSD) following cerebellar tumor resection has attracted much scientific attention. Taking into account the low frequency of occurrence of this syndrome – approximately 75 cases have now been described all over the world – the number of review studies [2-7] is remarkable. They all attempt to unravel the underlying mechanism and risk factors. The studies presented in this thesis also focus on these aspects. This clinical and scientific interest may be due to several circumstances:
1) MSD occurs in the acute postoperative course, while the child is under intensive and multidisciplinary care. Its occurrence is in dramatic contrast with the 'successful' resection of the tumor. The careful multidisciplinary analysis of MSD is of clinical value, as it contributes to a better understanding of the consequences of neurosurgery. Operative techniques, involving the possible occurrence of MSD have now been identified [8,9]. Clinical analysis has also revealed prognostic factors related to the duration of the mute phase and the moment of speech onset. The possibility of this complication is now well recognized and pointed out to the (older) child and the parents preoperatively.
2) The study of MSD is in line with recent clinical and research reports, focusing on other functions of the cerebellum than its control function during motor activity as well as challenging traditional concepts of cerebellar function [10-15].
Our neuropsychological findings are discussed in the light of current knowledge on cerebellar function and clinico-anatomic relations.

8.2. MSD is a pediatric syndrome

MSD has rarely been reported in adults [3]. Several explanations have been offered. The higher incidence in children was thought to be due te the influence of an emotional factor [4], playing a more crucial role in children than in adults. On the basis of our observations we conclude that a psychic factor cannot sufficiently explain the speechlessness, as the children were cooperative and cheerful during the mute phase when carrying out nonverbal tasks (Chapter 5). Moreover, it does not account for the following motor speech disorder.

Neuropathological explanations should be considered. A developmental perspective is offered by Jernigan and Tallal [16]. Analyzing morphological changes of the cerebellum in children and young adults, they report neocerebellar growth in late childhood and early adulthood, suggesting that damage to the cerebellum in this age range interferes with late maturation. This vulnerability to damage in childhood however does not explain why 80% of children with cerebellar tumors do not develop MSD.

It was possibe to elucidate specific risk factors associated with MSD in children (Chapter 2). Tumor type is one of the risk factors linked with MSD, as MSD occurs more frequently after surgical removal of medulloblastoma than of any other type of

tumor. Cerebellar medulloblastoma is a tumor typical of infancy and childhood. Of all patients, 77% are under 19 years of age [17]. The tumors grow rapidly and most frequently affect the cerebellar vermis, from which they expand into the fourth ventricle or infiltrate the cerebellar hemispheres, the cerebellar peduncles, or the dorsal brainstem [9]. The tumor growth into these structures makes intensive neurosurgical manipulation necessary. The histopathology and tumor location are different from medulloblastomas in adults [18], which may explain, why MSD is rarely reported in adults [3].

8.3. Mutism in MSD is of apraxic-like nature

Cerebellar mutism differs from other forms of acquired neurological mutism in childhood (ANCM). Neuropsychological analysis of the causes underlying ANCM (Chapter 4) – lack of mental activity, aphasia, cortical deafness, and anarthria – has revealed, that these factors do not explain the cerebellar mutism. As our studies show, MSD shows a pattern of stages which reflect the dynamics of this speech disorder. The first phase is a (short) period of bulbar syndrome, manifesting itself in swallowing problems. The second phase is characterized by the restoration of bulbar function, manifesing itself in finely coordinated movements of mouth and tongue when chewing and swallowing. However, these simple orofacial movements can not be initiated on request. During the next phase, simple voluntary orofacial movements are restored. The restoration of learned complex voluntary movements coincides with the onset of speech. This condition resembles apraxia, which is defined as a disorder of higher order motor behavior, in which 'the putting together of the motor program and the running of motor plans in response to external influences and internal thought is defective' [19]. It cannot be accounted for either by weakness, incoordination, sensory loss, or by incomprehension of or inattention to commands [20].

Other studies on MSD have suggested that apraxia, the loss of the ability to perform learned movements, underlies mutism in MSD [21-23]. We are reluctant to label the orofacial impairments in MSD as orofacial apraxia or 'apraxia of the cranial musculature' [20] for the following reasons: 1) the characteristics of orofacial apraxia are based on observations in adults; 2) apraxia is attributed to pathology affecting the parietal and frontal cortical areas of the dominant speech hemisphere; 3) acquired apraxia in adults is characterized by inappropriate movements or inappropriate elements within a movement [24] rather than by a loss of initiation. Given the fact, that other impairments of what used to be called 'higher cortical functions' have been described after cerebellar damage [14,15], the term 'cerebellar orofacial apraxia' may be appropriate to designate the underlying mechanism.

8.4. The study of ACD is still in its infancy

The most conspicuous speech deficits in MSD are articulation problems and slow speech rate with occasional deficits in pitch and vocal quality. Articulation errors are consistent and predictable. We labeled this motor speech disorder dysarthria in accordance with Darley et al's [25] definition. In adults, dysarthria is distinguished from a different motor speech disorder, apraxia of speech, which is a selective impairment in the programming of articulation movements that becomes manifest in unpredictable errors, increasing with word length, rendering speech slow. Apraxia of speech has been described as an acquired pure motor speech disorder in adults, but to the best of our knowledge there are no reports of acquired pure apraxia of speech in children.

A review of the literature on acquired childhood dysarthria (ACD) showed that detailed analysis of ACD has rarely been carried out, and that pediatric studies frequently refer to the adult model of distinct dysarthria types [25].

This model of distinct dysarthria types in adults [25] has been put forward more than two decades ago, but it is still universally applied. However, due to major advances in neurology and neuroimaging, shortcomings of the model have now become evident. Patients were assigned to different diagnostic groups on the basis of a clinical diagnosis. The underlying structural pathology could only be inferred, as CT and MRI were not yet available. The different diagnostic groups therefore constitute heterogeneous groups. Clinicoanatomical correlations are dubious in the light of present standards.

As a first step towards a classification of ACD we attempted to seperate speech features in children with dysarthria due to brainstem or cerebellar tumors (Chapter 7). We could demonstrate that speech features differed from those in adults with cerebellar or brainstem lesions, which underlines that the validity of the adult model of distinct dysarthria types for ACD requires further investigation.

We are aware of major limitations of the study. The distinction between the groups may be somewhat artificial as cerebellar tumor resection may produce additional pathology, and brainstem tumors are sometimes exophytic with various extensions. We choose to analyze speech features on one occasion in each child. The changes over time in the cerebellar group had been presented elsewhere. In the brainstem group the irradiation and chemotherapy may temporarily alleviate the speech pathology, but this aspect was beyond the aim of the study.

We did not attempt to analyze clinicoanatomical correlations as the tumoral etiology does not enable to clarify which substructures contribute to the observed speech disorders. The study illustrates the state of the art and offers a starting point for further research.

8.5. MSD reflects the foreign policy of the cerebellum

Cortical – cerebellar pathways play an important role in speech motor control. Preparation and initiation of speech as well as control during cortically generated speech output is realized by pathways from the primary and premotor cortex via the

pons to the cerebellum, through dentato-thalamo-cortical pathways, and through corticobulbar and corticospinal tracts [27]. By the reciprocal connections within the cerebello-thalamo-cortical loop, structural cerebellar lesions may regulate neural activity, blood flow and metabolism in the thalamus and in cortical regions [28]. Consequently, cerebellar lesions may produce neuropsychological deficits which are usually attributed to cerebral cortical lesions and therefore designated as higher cortical functions [14,15]. Such a mechanism could account for our findings. The severe preoperative hydrocephalus is a risk factor which may lead to MSD. This suggests that the pressure of the severely increased ventricular size in combination with the trauma of the cerebellar surgery is a prerequisite for the shutting down of the cerebello-thalamo-cerebral circuit, thus leading to such symptoms as MSD and apraxia-like orofacial dysfunction. Since the abovementioned symptoms nearly always have a delayed onset and are always transient, there seems to be a functional disturbance within the circuitry rather than structural damage. This assumption is supported by the results of SPECT scans carried out in one child (Chapter 5, Case 1): transient hypoperfusion of both frontal cerebral lobes was observed, which suggests diaschisis [28] as the underlying pathophysiological mechanism. In the light of these clinical features and neuroimaging findings we consider MSD not an exclusively cerebellar syndrome.

New techniques in (functional) neuroimaging will be a major contribution to a better understanding of clinico-anatomo-physiological correlations. Consequently, strict localizationistic concepts in neuropsychology will be replaced by models illustrating the teamwork within functional systems.

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9

Nederlandse samenvatting

Dit proefschrift omvat neuropsychologische studies rondom een syndroom, dat kan optreden bij kinderen na een chirurgische ingreep, waarbij een tumor uit de kleine hersenen – cerebellum – wordt verwijderd. Het syndroom is in 1986 voor het eerst beschreven en sindsdien zijn ongeveer 75 gevalsbeschrijvingen gepubliceerd. Het komt voor bij ongeveer 20% van de kinderen, die een dergelijke operatie ondergaan. Na de operatie ontwaken de kinderen uit de narcose en beginnen te praten. In de volgende uren tot dagen verliezen zij het vermogen tot spreken, aangeduid als mutisme. Deze mutistische fase kan in duur variëren van enkele uren tot maanden, maar is van voorbijgaande aard. Wanneer de kinderen weer gaan spreken, hebben ze een spraakstoornis, die dysarthrie genoemd wordt. Deze dramatische postoperatieve complicatie na verwijdering van een cerebellaire tumor – aangeduid als MSD (mutism and subsequent dysarthria) – is vrijwel uitsluitend bij kinderen geobserveerd. In dit proefschrift wordt nader ingegaan op de twee aspecten van dit syndroom, mutisme en dysarthrie.

In *Hoofdstuk 2* en *Hoofdstuk 3* worden de risicofactoren voor het ontstaan van dit syndroom beschreven. De onderzoeksgroep bestaat uit 44 kinderen, die in het Academisch Ziekenhuis Rotterdam aan een cerebellaire tumor werden geopereerd, waarvan 12 kinderen MSD vertoonden. Bij een vergelijking tussen kinderen met en zonder MSD blijkt, dat er verschillen bestaan in plaats en aard van de tumor, en bijkomende neurologische complicaties door de tumor en de operatie. De circulatie van hersenvocht kan door de tumorgroei verstoord zijn, waardoor holtes met hersenvocht – ventrikels – vergroot worden – hydrocephalus – en druk uitoefenen op omliggende gebieden. In die gevallen wordt preoperatief een shunt aangelegd. De groei van de tumor vanuit het cerebellum in de hersenstam – meestal optredend bij een bepaald type tumor (medulloblastoom) –, maakt intensieve manipulatie door de neurochirurg bij de verwijdering van de tumor noodzakelijk. Dit zijn belangrijke risicofactoren voor het ontstaan van MSD. Op basis van deze risicofactoren moet ervan uitgegaan worden, dat MSD niet uitsluitend het gevolg is van het structurele letsel in het cerebellum, en dat de verstoorde functie van andere gebieden, nauw verbonden met het cerebellum, bijdraagt aan het ontstaan van MSD.

Verworven mutisme bij kinderen kan optreden bij zeer uiteenlopende aandoeningen in het centraal zenuwstelsel. De verschillen in etiologie en localisatie van de beschadiging maken mutisme tot een zeer heterogeen beeld met vele gezichten. Deze worden in *Hoofdstuk 4* beschreven. Volgens een standaardprotocol beoordeelden we het gedrag van vier mutistische kinderen, waarvan één kind met MSD, op factoren, die bij het spreken een rol spelen: helderheid en alertheid, motorische stoornissen van romp en ledematen, mondmotoriek, het vermogen om geluid voort te brengen, communicatiebehoefte, taalbegrip, en emotionele aspecten. Deze gedetailleerde analyse van gedragskenmerken bij mutistische kinderen maakt het mogelijk een uitspraak te doen over de onderliggende oorzaak en de herstelkansen. MSD onderscheidt zich van mutisme ten gevolge van letsels in andere hersengebieden.

De literatuur over MSD bevat vele hypothesen over het mechanisme dat aan het mutisme ten grondslag ligt. Emotionele aspecten – het kind is angstig door de

omgeving van de intensieve zorg afdeling, de vele vreemde gezichten om zich heen en de pijn – werden van belang geacht als oorzaak voor het mutisme, maar in de meeste studies wordt een vermoeden geuit, dat er problemen bestaan rondom de mondmotorische bewegingen, die voor het spreken van belang zijn. In eerder onderzoek (Hoofdstuk 2) was het opgevallen, dat kinderen in de mutistische fase niet in staat waren op verzoek mondbewegingen te maken. In *Hoofdstuk 5* werd daarom de mondmotoriek van vijf kinderen met MSD zorgvuldig geanalyseerd. Uit deze analyse gedurende de mutistische fase en kort na de terugkeer van de spraak blijkt het volgende: tijdens de mutistische fase zijn kinderen met MSD niet in staat simpele orofaciale bewegingen – ogen sluiten, kaken openen en sluiten, lippen aflikken – op verzoek te maken of te imiteren, terwijl deze bewegingen tijdens kauwen, slikken en drinken ongestoord zijn. Uit de analyse blijkt verder, dat het herstel van het vermogen om op verzoek complexe – doe-alsof – bewegingen te maken, samenvalt met de terugkeer van de spraak. Deze discrepantie tussen de onwillekeurige en de willekeurige bewegingen – apraxie – wordt daarom beschouwd als de oorzaak van het mutisme, temeer daar andere factoren, die een rol spelen in andere vormen van mutisme, zoals verlammingen, een taalstoornis, of emotionele factoren uitgesloten konden worden.
Na de mutistische fase hebben MSD kinderen een ernstige motorische spraakstoornis – dysarthrie –, maar deze herstelt meestal in een periode van enkele maanden. Op basis van de primaire localisatie van de tumor wordt deze spraakstoornis in de literatuur meestal aangeduid als cerebellair. Daarbij wordt verwezen naar de kenmerken van dysarthrie bij volwassen patiënten met cerebellair letsel, waarbij de cerebellaire dysarthrie kenmerken zou vertonen, die lijken op stoornissen van de overige motoriek – ataxie – staccato spraak met onvoorspelbare uitschieters in de articulatie. Het was echter onduidelijk of dit 'volwassen model' wel toegepast mag worden op kinderen.

Hoofdstuk 6 geeft daarom een overzicht van alle beschrijvingen van verworven dysarthrie bij kinderen. Gevalsbeschrijvingen werden geclassificeerd op basis van neuroradiologische gegevens omtrent plaats van het letsel en de motorische stoornis. Uit dit overzicht blijkt, dat de kennis over dysarthrie bij kinderen is gebaseerd op een beperkt aantal studies, waarvan de dysarthrie na cerebellaire tumor resectie het meeste aandacht heeft gekregen. De spraakkenmerken van de dysarthrie worden zelden gedetailleerd beschreven. Duidelijke overeenkomsten met het volwassen model komen niet naar voren, met uitzondering van de dysarthrie ten gevolge van beschadigingen in de basale gangliën. Op basis van dit literatuuronderzoek concluderen wij, dat er onvoldoende bewijs is dat het volwassen model van dysarthrietypen op kinderen van toepassing is, en dat verworven dysarthrie bij kinderen een eigen classificatie vereist.

Een eerste stap tot een dergelijke classificatie vormt het onderzoek, dat in *Hoofdstuk 7* beschreven wordt. Dit onderzoek heeft betrekking op een analyse van dysarthrische spraakkenmerken bij twee groepen kinderen met eenzelfde etiologie – tumor – maar een verschillende localisatie – cerebellum of hersenstam –. In beide groepen komen ataxie, pyramidebaan verschijnselen en dysfuncties van de hersenzenuwen voor. In

beide groepen treden frequent articulatie problemen op, maar er zijn ook opmerkelijke verschillen. In de cerebellaire groep is er sprake van een traag spreektempo. Echter, de staccato spraak en het onvoorspelbare articulatiefalen, dat karakteristiek is voor volwassenen met cerebellaire aandoeningen, wordt niet gevonden. De kinderen met hersenstamtumoren vertonen een opvallende hypernasale spraak, die ook bij volwassenen met hersenstamletsel is beschreven. Er wordt geen eenduidige relatie gevonden tussen de spraakkenmerken en de stoornissen in de overige motoriek. We concluderen, dat het volwassen model van typen dysarthrie, waarbij de motorische stoornis verondersteld wordt zich te weerspiegelen in de spraak, niet zonder meer toegepast mag worden op kinderen met tumoren. De kennis over de verworven kinderdysarthrie staat nog in de kinderschoenen, waarbij dit onderzoek een eerste stap is om te komen tot een eigen classificatie van verworven dysarthrie bij kinderen.

10 Summary

This thesis comprises neuropsychological studies on a syndrome which may occur after the surgical resection of a cerebellar tumor. The first description of the syndrome dates from 1986 and until 1997, 75 cases studies have been published. It is estimated that the syndrome occurs in approximately 20% of the children who underwent cerebellar tumor resection.
After the operation the children awake from anesthesia and start to speak. In the next few hours or days they loose the ability to speak, which is designated as the mute phase. This mute phase is of variable duration and may last hours up to several months, but is of a transient nature. At speech onset, the children have a motor speech disorder which is labeled as dysarthria. This dramatic postoperative complication after cerebellar tumor resection – designated as MSD (mutism and subsequent dysarthria) – has rarely been observed in adults. The studies presented in this thesis focus on the two aspects of the syndrome i.e. mutism and dysarthria.

In *Chapter 2* and *Chapter 3* risk factors for the development of the syndrome are described. The patient group includes 44 children, who underwent cerebellar tumor resection at the University Hospital Rotterdam, of whom 12 children suffered from MSD. A comparison of the children with MSD and those without MSD shows that there are differences with respect to localization, type of the tumor, and additional neurological deficits associated with tumor type and surgical procedure. Tumor growth may obstruct the circulation of cerebrospinal fluid leading to the enlargement of chambers filled with cerebrospinal fluid – ventricles – , which may lead to pressure on the surrounding tissues. In those cases preoperative shunting is required. The growth of the cerebellar tumor into the adjacent brainstem – frequently occurring with a specific type of tumor (medulloblastoma) – necessitates the neurosurgeon to manipulate the brainstem while resecting the tumor. On the basis of these risk factors it is reasonable to conclude that MSD is not exclusively the result of structural damage to the cerebellum, and that additional impairment of areas, which are functionally related to the cerebellum, contributes to the development of MSD.

Acquired neurological mutism in childhood (ANCM) may be the result of various diseases of the central nervous system. Due to differences in etiology and localization of the damage, mutism is a heterogeneous clinical picture with many faces. The various faces of ANCM are described in Chapter 4. Four mute children, of whom one suffered from MSD, were examined by means of a standard protocol, comprising behavioral examination of factors, which are known to play a role for speech: mental activity and alertness, motor impairment of trunk and limbs, orofacial musculature, phonation, communicative urge, language comprehension and emotional aspects. This detailed analysis of behavioral features of mute children allowed us to indentify the underlying mechanisms of ANCM and to make prognostic statements as to the recovery from mutism. MSD constitutes a type of mutism different from forms of mutism caused by impairments in other areas of the brain.

The literature on MSD has put forward hypotheses on the underlying mechanism. Emotional factors – the child is frightened by the surroundings and the hospital staff

of the intensive care unit – were thought to be of importance. However most studies refute these emotional factors as a cause of MSD and propose that orofacial movements required for speech are impaired. From our research presented in the Chapters 2 and 4 we got the impression that children with MSD were unable to perform orofacial movements on request.
We therefore analyzed orofacial movements in five children with MSD which study is presented in Chapter 5. This analysis during the mute phase and shortly after the recurrence of speech revealed the following: during the mute phase children are unable to perform or to imitate simple orofacial movements on request – eye closure, jaw opening and closure, licking off the lips – while these movements occur automatically during the acts of chewing and swallowing or drinking. The analysis further revealed that the restoration of the ability to perform complex orofacial movements coincides with the onset of speech. The discrepancy between automatic and voluntary movements – apraxia – therefore is considered the cause of mutism in MSD, so much the more as other factors which may play a role in other types of mutism – pareses, language disorder, or emotional factors – could be ruled out.

In MSD the mute phase is followed by a period of motor speech disorder – dysarthria –. Initially the dysarthria is severe but it usually recovers within a few months. On the basis of the primary site of the tumor, reports in the literature designate this speech disorder as cerebellar, thereby implicitly referring to speech features of adult patients with cerebellar lesions. The adult cerebellar dysarthria is thought to reveal speech features which resemble the accompanying motor impairment – ataxia – : staccato, scanning speech with irregular breakdown of articulation. It remained unclear, whether the adult model could be applied to children.
Chapter 6 therefore presents a review of the literature on acquired childhood dysarthria (ACD). Case studies were classified on the basis of neuroradiological evidence of the lesion site and on the basis of the motor impairment. The review shows that knowledge on ACD is based on a limited number of studies mostly pertaining to dysarthria following cerebellar tumor resection. Dysarthric speech features are rarely reported in detail. Similarities with the adult model of distinct dysarthria types rarely emerge except for dysarthria following basal ganglia lesions. On the basis of the literature review we conclude that there is insufficient evidence that the adult model of distinct dysarthria types is valid for ACD and that ACD needs its own classification.

As a first step towards such a classification we carried out a study which is presented in Chapter 7. We compared speech features in two groups of children: they all became dysarthric due to the same etiology – tumor – but site of the tumors was different – cerebellum or brainstem –. In both groups ataxia, pyramidal signs, and cranial nerve deficits were frequent. Both groups revealed articulation problems but the analyses also revealed striking differences. The dysarthria in the cerebellar group was characterized by slow speech rate. However, scanning speech and irregular articulatory breakdown, characteristic of cerebellar dysarthria in adults, was not observed. Children with brainstem tumors revealed hypernasality, which has also been observed in adults with brainstem lesions. We could not identify a clearcut

correlation between speech deficits and motor impairment of trunk and limbs. From our observations we conclude that the adult model of speech features, mirroring motor disorder, may not be simply applied to children with dysarthria caused by tumors.

In general we conclude that the knowledge on ACD is still in its infancy.

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Curriculum Vitae

Marijke van Mourik werd geboren op 18 mei 1953 te Velp, Gelderland. Na de lagere school te Velp bezocht zij het Stedelijk Gymnasium te Arnhem en het Wagenings Lyceum, waar zij in 1971 het eindexamen Gymnasium α behaalde. Zij studeerde Psychologie aan de Rijksuniversiteit Utrecht, waar zij in 1979 afstudeerde in de Funktieleer, Cognitieve Functiestoornissen. Van 1977 tot 1980 was zij werkzaam in de Neurologische Kliniek 'August Bier' te Malente-Gremsmühlen, Duitsland.
Zij verrichtte neuropsychologisch onderzoek bij kinderen en volwassenen met traumatisch hersenletsel, cerebrovasculaire aandoeningen en hersentumoren. Van 1980 tot 1990 was zij verbonden aan de Afdeling Neuropsychologie van de Medische Faculteit Rotterdam en in dienst van de Stichting Afasie Rotterdam. In die periode was zij betrokken bij het opzetten van diagnostiek en therapeutische richtlijnen bij afasiepatiënten. Tezamen met Drs. M. Koning-Haanstra publiceerde zij het Leerboek 'Afasie, een multidisciplinaire benadering', en met Drs. F. van Harskamp publiceerde zij onderzoek naar behandelingsmogelijkheden bij patiënten met een globale afasie. Onder supervisie van Dr. H.R. van Dongen werd zij in 1985 geregistreerd als Klinisch Psycholoog. Sinds 1989 is zij in dienst van het Ziekenhuis Walcheren.
Zij is werkzaam als Klinisch Psycholoog en Afdelingshoofd van de Medische Psychologie, en heeft als belangrijkste aandachtsgebieden de neuropsychologie, de diagnostiek bij kinderen met taal- en spraakachterstanden en de pychosociale zorg voor oncologiepatiënten. Zij is moeder van drie kinderen.