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Transient Mutism and Speech Disorders After Posterior Fossa Surgery in Children with Brain Tumours

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Summary

Four patients aged 5 to 9 years with large tumours located in the posterior fossa (PNET, ependymoma or astrocytoma) are presented. Patients received standard neuropsychological assessments, including speech evaluation, prior to surgery. Following tumour resection, these 4 children developed transient mutism or different types of speech and cognitive disorders, associated with behavioural disturbances.

We describe course and results of repeated postoperative neurological and neuropsychological evaluations. Full recovery of speech was seen in 3 out of 4 patients; the only child with persistent symptoms was the one who already had neuropsychological deficits before surgery. However, despite fast recovery of the speech disorders more persistent behavioural problems were found in 3 out of 4 patients.

Possible pathogenesis and anatomical location of this "cerebellar speech syndrome" are discussed, as well as the relevance of repeated neuropsychological assessments.

Keywords: Posterior fossa surgery; brain tumour; children; speech disorder.

Introduction

Mutism, defined as the complete absence of speech in a subject with unimpaired consciousness, may result from psychiatric disease (i.e., functional or non-organic mutism) or from organic lesions of the nervous system. The latter can be divided into several subtypes:

1) Complete Broca's aphasia due to a lesion of the motor cortex of the dominant hemisphere.

2) Akinetic mutism associated with lesions of the mesencephalon.

3) Pseudobulbar palsy with diffuse bilateral hemispheric lesions.

4) Mutism following bilateral thalamotomy.

5) Bilateral pharyngeal or vocal cord paralysis associated with peripheral nerve lesions[10, 17].

More recently, a "cerebellar" form of mutism, following posterior fossa (PF) surgery has been described in both adults and children[2, 8, 10, 12, 13, 17, 21]. Such cerebellar muteness represents a distinct clinical syndrome and is characterized by:

1) a postoperative period of latency ranging from 18–72 hours during which the patient can speak,

2) the fact that the mutism cannot be explained by additional postoperative deficits of the cranial nerves, long tracts or peripheral speech organs.

This mutism syndrome has been considered as either the ultimate form of cerebellar dysarthria, namely anarthria, or as an apraxia of speech. The majority of patients demonstrated complete mutism but others made sounds, wept or whined. Subgroups of children were not totally mute but showed a marked decline in speech, saying only "yes" or "no" when asked questions. The mutism was transient in all cases; recovery varying from weeks to months. Almost all patients passed through a phase of cerebellar or ataxic dysarthria when they gradually regained speech. Many patients were apathetic during the mutistic phase, with some of them showing adequate nonverbal reactions while others failed to do so. Unfortunately, pre-operative assessment of speech and language functions was rarely available in those children with brain tumours who presented with a mutism syndrome. Hence, damage caused by the tumour and by the surgery could not fully be distinguished.

We here report 4 patients who recently demonstrated varying types of cerebellar speech disorders following the surgical removal of a PF tumour. These children were part of a prospective and longitudinal study on the late-effects of neuropsychological assess-

Table 1. *Clinical Characteristics of 4 Patients with Posterior Fossa Tumours at Diagnosis*

Patient	Age[a]/ sex	Tumour site and size	Histology	Neuropsychological assessment
1	5,3 M	vermis, 4th ventricle 4 cm	pilocytic astrocytoma	normal speech fluency[b] score: 16 (above average) IQ[c]: above average no cognitive deficits
2	5,7 M	vermis, 4th ventricle 4,5 cm	PNET	normal speech fluency score: 17 (above average) IQ: average no cognitive deficits
3	9,5 M	vermis, 4th ventricle 4,5 cm	ependymoma	decline in spontaneous speech; monotous, bradyfrasia, dysarthria fluency score: 7 (below agerage) IQ: below average various cognitive deficits[d] change in personality (anamnesis)
4	9,2 M	vermis, 4th ventricle 4 cm	PNET	normal speech fluency score: 22 (above average) IQ: above average no cognitive deficits change in personality (anamnesis)

[a] in years, months.
[b] fluency score: number of animals enumerated in 1 minute[19].
[c] IQ: estimated on 6/10 subtests of Wechsler scales (WPPSI or WISC-R)[22, 23].
[d] Normal cognitive functions until 1 year before diagnosis.

ment of brain tumour treatment; they all had had neuro-psychological assessments, including speech evaluation, prior to surgery[15]. Diagnosis and further evaluation of the postoperative speech disorder appeared highly important concerning optimal patient care. Despite good recovery of speech functions, 3/4 patients suffered from other persistent neuropsychological deficits.

Case Material and Methods

Neuropsychological evaluation prior to and after surgery included measures of intelligence, auditory and visual memory, attention, speed, visuo-motor-constructive functions, speech and language development, global school achievement and fine-motor functioning[3, 5, 7, 9, 14, 16, 19, 20, 22, 23]. Brainstem auditory evoked potentials (BAEP) including stimulation with both positive and negative clicks (1600 ×) were only examined in patient 1. The characteristics of patients are shown in Table 1.

Case 1

This 5 year old righthanded boy had non-specific health problems for one month with progressive headaches and vomiting. He presented with strabismus, nystagmus, choked discs, sixth nerve palsy and global ataxia. CT scanning revealed a large cerebellar tumour associated with obstructive hydrocephalus. He received an external ventricular drain (EVD) on the day of admission; on day 4 he was lucid and co-operative and neuropsychological assessment showed no deficits, with undisturbed speech and language functions, and normal behaviour.

On day 8, a standard suboccipital craniectomy was performed. After opening the dura, part of the tumour presented between the herniated tonsils. The tumour – a pilocytic astrocytoma – was removed subtotally, following partial splitting of the vermis. A thin layer of tumour, adherent to the floor of the fourth ventricle, had to be left in situ.

Postoperative course: On the first postoperative day he said 2 sentences but was totally mute thereafter for 4 weeks. He did not utter any sounds, was very apathetic and did not use nonverbal communication. External verbal stimuli did not seem to "get through". BAEP examination showed a clear increase in the ipsilateral III–V and I–V interpeak latencies, both right and left, indicating a central conductive disorder at high pontine level.

Neurological examination showed slight left sided facial paresis, left sided hemiparesis and inability to swallow.

In the 5th postoperative week, his first words were "no", "yes", "mom" and "dad". His speech was slow, weak, dysarthric and monosyllabic at first. He gradually regained his former speech and 6 months post surgery neuropsychological assessment showed almost normal speech functions with very slight dysarthria. Neurologically, the cranial nerve paresis had disappeared completely and motor function was almost symmetrical.

However, he suffered from a serious attention deficit and behaviour disorder. One year after surgery the boy lives still in a day care centre for brain-injured children. His speech has returned to a normal, pre-morbid level.

Case 2

Another 5 year old left-handed boy had a 2 months history of early morning vomiting and headaches. On admission to our hospital he showed choked discs and a slight sixth nerve palsy on the left. CT scanning revealed a cerebellar tumour with obstructive hydrocephalus and he immediately received an EVD. Three days later he was alert and co-operative; neuropsychological assessment showed normal cognitive functions with no speech or language deficits and normal behaviour.

On day 5, a standard suboccipital craniectomy was performed. The tumour – PNET – was removed except for some fragments adherent to the brainstem. A temporary intra-operative complication, consisting of tachycardia and haemodynamic instability was managed adequately.

Postoperative course: He was intubated for 4 days. On the 5th postoperative day he said one spontaneous word (repeating the word "perfume" when seeing the bottle) and another word on day 7 (mentioning the name of the principal character in a children's story). Otherwise, he was mute and apathetic for 2 weeks, with occasional crying and uttering of sounds. He did not try to communicate nonverbally but sometimes he could follow simple commands. Neurologically, there was a slight left sided paresis, particularly of his arm, but no signs of lower cranial nerve dysfunction. Two weeks after operation he started talking by saying "no", calling his sister's name and counting up to 12. He rapidly regained speech and spoke fluently in sentences 4 weeks after the surgery. He never was dysarthric but his speech was a bit lisping. In his spontaneous speech some verbal paraphasias were noted. The fluency-test (number of animals enumerated in one minute) yielded many perseverations. He also demonstrated a marked change in behaviour which had become unpredictable, perseverative, inappropriate and poorly controlled. For one week, he stuffed his mouth full of food and uneatable things.

A course of 35 Gy cranial irradiation with 53 Gy tumourboost was started. Neuropsychological assessment 6 months after surgery showed significant improvement in behaviour and cognitive functions. Speech has returned to normal with an occasional paraphasic error. Neurological symptoms and signs were remarkably improved with only slight residual left sixth nerve palsy and minimal ataxia. One year post-surgery he is still slow in school with a poor sustained attention. We have not yet decided whether he can stay in his regular class or should be referred for special education.

Case 3

This 9 year old, righthanded boy presented with a long history of decline in school performance. He could not follow his class's space, showed impaired handwriting and became withdrawn. Occasional vomiting started 8 months and atactic gait 1 month before admission. Repeated psychological consultations elsewhere considered emotional disorder because of tragic family circumstances.

On admission, an MRI revealed a cerebellar tumour with no signs of hydrocephalus. Neurological examination showed slight papilloedema, hearing loss of the left ear and ataxia, predominantly of the right arm. Neuropsychological assessment showed an IQ of

Table 2. *Evaluation of Expressive Speech in 4 Patients After Surgical Removal of a Posterior Fossa Tumour*

Patient	1 week post-surgery	5 weeks post-surgery	1 year post-surgery
1	mutism; latency 24 hours fluency score[a]: 0 no reaction to commands	some spontaneous, monosyllabic speech bradyfrasia dysarthria fluency score: 9	normal speech no dysarthria fluency score 13 (average) serious behaviour- and attention-deficit disorder, therefore special education
2	mutism; latency? fluency score: 0 some reaction to simple commands	fluent spontaneous speech with some semantic-aphasic verbal paraphasias no dysarthria slight lisping fluency score: 4	normal speech fluency score: 10 (average) poor sustained attention; regular education?
3	no spontaneous speech "yes", "no", "okay" in reply to questions fluency score: 0 could perform commands	good spontaneous speech but simple and short sentences bradysfraisa dysarthria fluency score: 9	normal speech except for bradyfrasia no dysarthria fluency score: 13 (average) various cognitive deficits; special education
4	no spontaneous speech "yes", "no", in reply to questions fluency score: 1 could perform commands	little spontaneous monosyllabic speech slight dysarthria fluency score: 10	normal speech no dysarthria fluency score 21 (above average) normal school performance slight change in personality

[a] Fluency score: number of animals enumerated in 1 minute[19].

73, very poor verbal-auditory memory, lethargy, bradyfrenia and bradyfrasia. There was little spontaneous and dysarthric speech but no signs of language disorder. His parents told us that his school performance and emotional development was up to standard until 1 year ago.

At surgery, the tumour, an ependymoma, was totally removed from the fourth ventricle by a standard suboccipital craniectomy.

Postoperative course: There was hardly any spontaneous speech in the first postoperative weeks. He would only say "yes", "no", "okay" if asked questions. However, he walked around and could perform commands. Neurologically, he showed an increased right-sided ataxia, which mainly resolved within weeks. After 4 weeks he spoke very slowly, in short and simple but proper sentences with a slight dysarthria. He received a course of 68 Gy local irradiation. Six months after operation there was still improvement in verbal expression and other cognitive functions but both the mental and motor impairment required special education for handicapped children. There was still some improvement one year after surgery but the boy persistently needs special education because of the (pre-existent) bradyfrenia, bradyfrasia, poor memory and ataxia of the dominant hand.

Case 4

Another 9 year old, righthanded boy had a year long history of negative personality changes and decline in fine motor performance, as demonstrated in poor handwriting and difficulties in playing the piano. Three months before admission, he started to vomit; 2 months later followed by headaches, double vision and ataxtic gait. At admission, an MRI showed a cerebellar tumour with obstructive hydrocephalus. Neurological examination demonstrated papilloedema and a slight left sixth nerve palsy.

He immediately received an EVD and neuropsychological assessment was performed on day 4. No cognitive deficits and no speech or language disabilities were found.

On day 5, the tumour (PNET) was totally removed through a suboccipital craniectomy, after splitting the caudal part of the vermis. The postoperative course was complicated by ventriculitis; the external drain was removed 3 weeks postoperatively.

At postoperative neurological examination he showed a more pronounced ataxia and sixth nerve palsy, both of which improved in the following weeks. Except for "yes" and "no", which were often misused, no spontanous speech was heard during the first 3 weeks. He was very apathetic. Slowly he started to use occasional one-word sentences if he wanted something, such as "milk" or "t.v.". His articulation was slightly dysarthric. His behaviour was negative, obstructive and impertinent and very different from his premorbid personality. In this case the patient gave the impression that he also did not want to talk. Neuropsychological assessment 6 months post-surgery showed no impairment compared to the pre-operative assessment, with good language and speech functions. However, his parents noted occasional verbal paraphasias in this spontaneous speech, such as book for paper, which he had never shown before. Neurologically only minimal nystagmus remained. He returned to his former school with good results but one year after surgery his behaviour is still slightly different, i.e. irritable, compared to his earlier personality.

Postoperative course and evaluation of speech functioning for the 4 patients is described in Table 2.

Discussion

The patients presented here not only represent examples of pure mutism following PF surgery. They include cases of various speech disorders with or without dysarthria, ranging from true mutism in patients 1 and 2 to a marked decline in speech in patients 3 and 4. Therefore, we prefer to call this phenomenon "cerebellar speech disorder".

In patient 1, no reaction, neither verbal nor non-verbal, could be provoked. This boy gave the impression that external verbal stimuli were not processed well, as was confirmed by BAEP examination. In patient 2, the period of latency is unknown because he was intubated during the first postoperative days. He did not speak for 10 days thereafter except for 2 occasional words, suggesting that he could speak but had not the drive to do so. In patient 3, dysarthria and cognitive deficits were already present at diagnosis but these symptoms and signs increased post-operatively. He never was totally mute but showed a marked decline in spontaneous speech with bradyfrenia and bradyfrasia. Patient 4 showed the best and fastest recovery with hardly any neuropsychological abnormalities 6 months after surgery. In this case, the patient gave the impression that he deliberately did not want to speak. Hence, recovery of speech was seen in 3/4 patients, the only patient with persistent bradyfrasia was the patient who already had deficits prior to surgery.

Although the decline in speech was the most obvious and problematical symptom in these children, there certainly was another brain disorder, that is, a general lack of drive. All children showed apathy during the first post-operative weeks. This lack of drive which turns to a behavioural disorder, reflecting deficits in control functions, is also noted after diffuse head injury in children[4].

The pathogenesis and anatomical location of this cerebellar speech and behaviour disorder are still unclear. Most cases in the literature had lesions of the brainstem and/or cerebellar vermis, with or without involvement of one or both cerebellar hemispheres. Extensive destruction of the (para) median, often bilateral, cerebellar structures including vermis, cortex, peduncles, and deep nuclei, is probably the most important anatomical substrate. The frequent dysarthria following the period of mutism also strongly suggests that cerebellar damage is the decisive factor underlying this syndrome. This also explains why these speech disorders are not seen after surgery for supratentorial tumours. Common factors in our patients included a

large midline tumour and involvement of the fourth ventricle resulting in hydrocephalus and pressure on the brainstem, and sixth nerve palsy on the left side. Additional aetiological factors, not restricted to the PF, include transient vascular disturbances of the brainstem due to ischaemia or oedema and disturbances of CSF circulation. The 24–72 hours delay after the operation, during which the patient can speak, also argues for a circulation or vascular aetiology, including post-operative swelling of the brain.

The lack of drive followed by cognitive and behavioural disorders with poor stimulus control could be explained by a global diencephalic dysfunction.

However, there is also growing evidence that the cerebellum contributes to verbal functioning, including verbal intelligence, verbal learning and fluency[1]. Positron emission tomography (PET) studies also have suggested that the cerebellum participates in many non-motor processes[11].

The question why the phenomenon of cerebellar speech disorder occurs predominantly in children, has not yet been solved.

We strongly recommend repeated neuropsychological assessments if a patient does not speak following PF surgery, for several reasons. First of all, this cerebral-organic syndrome must be distinguished from a psychogenic aetiology of non-speaking as was considered by Volcan *et al.* in one of their patients[21]. After the diagnosis "cerebellar speech disorder" has been established, results of the neuropsychological evaluation can be used for referral to a speech therapist and to advise parents and hospital staff with regard to the communication with the child and to explain possible changes in behaviour. Particularly deficits in language comprehension, as in our patients 1 and 2 are often not recognized or underrated. Finally, repeated assessments can determine degree of recovery and possibilities of school re-entry. If no (neuro)psychologist or speech therapist is available, recording of speech and simple tests as picture/object naming, writing, drawing and reading, word fluency, repeating sentences, can be applied by physician or nurse. Naturally, complete removal of a brain tumour has the highest priority as the first part of successful brain-tumour treatment but lowering of morbidity is a second important aim. Although the mechanism of this speech disorder is not fully understood, neuropsychological assessment can have an important contribution in the best management of these paediatric brain tumour patients.

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