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Posterior fossa syndrome in adults: A new case and comprehensive survey of the literature

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ABSTRACT

Although the posterior fossa syndrome (PFS) can be considered as an aetiologically heterogeneous condition affecting children and adults, it most often occurs in paediatric patients after cerebellar tumour surgery. In patients with a tumoural aetiology, the syndrome is typically characterised by a short symptom-free postoperative period followed by mutism of variable duration and behavioural and affective changes. More than 200 paediatric cases have been described but reports of adult patients are extremely rare.

This paper discusses PFS in adults on the basis of a comprehensive literature survey and describes the pre- and postoperative findings in a new adult patient. In the preoperative phase, cognitive, behavioural and affective abnormalities were identified, matching a diagnosis of cerebellar cognitive affective syndrome (CCAS) (Schmahmann and Sherman, 1998; Schmahmann, 2004). The immediate postoperative course was characterised by prefrontal-like behavioural and affective abnormalities, peduncular hallucinations and confusion evolving to psychosis. Akinetic mutism subsequently developed, lasted for 12 days and then alternated with episodes of diminished responsiveness in which pathological laughing and crying (PLC) occurred. Akinetic mutism resolved after treatment with a non-ergoline dopamine-agonist but CCAS persisted during longitudinal follow-up.

From a semiological point of view "relapsing-remitting akinetic mutism" and PLC in our patient might add relevant information to current insights in the clinical expression of the PFS. As evidenced by a close parallelism between single photon emission computed tomography (SPECT) and clinical findings, CCAS as well as PFS seem to reflect functional disruption of the cerebello-cerebral network involved in cognitive, behavioural and affective functions. These findings may indicate that both syndromes share overt semiological resemblances and a common pathophysiological substrate. Consequently, CCAS and PFS may both be regarded as cerebellar-induced clinical conditions showing different aspects of a spectrum that range in degree of severity and symptom duration.

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1. Introduction

The posterior fossa syndrome (PFS) is characterised by various linguistic, cognitive and behavioural-affective symptoms which may develop after aetiologically different lesions of the cerebellum. Although the condition is typically found in children after surgery of a cerebellar tumour, it has been reported in children after trauma (Ersahin et al., 1997; Fujisawa et al., 2005), stroke (Sinha et al., 1998; Baillieux et al., 2007), or infection (Drost et al., 2000; Riva, 1998).

In children with a posterior fossa tumour, the syndrome typically develops after a short interval of relatively normal postoperative functioning, which may last from a few hours to a couple of days. After this symptom-free period, cerebellar mutism may become evident which may last for one day to several months (Levisohn et al., 2000; De Smet et al., 2007). Mutism is generally accompanied by frontal-like neurobehavioural abnormalities such as apathy or reduced initiative, unconcern and whining. Although the loss of speech has been generally considered as the hallmark feature of the PFS, a wide range of postoperative neurobehavioural defects have been found in the absence of mutism: decreased initiation of voluntary movements (e.g., Siffert et al., 2000), eye-lid apraxia (e.g., Pollack, 2001), executive dysfunction (e.g., Levisohn et al., 2000; De Smet et al., 2009), poor problem-solving (Pollack et al., 1995; Clerico et al., 2002), mnestic disorders (Kingma et al., 1994; Humphreys, 1989), reduced attention-span (Kingma et al., 1994; Aarsen et al., 2004), visual-constructive deficits (Riva, 1998; De Smet et al., 2009), and symptoms consistent with cerebellar cognitive affective syndrome (CCAS) (Schmahmann and Sherman, 1998; Catsman-Berrevoets and Aarsen, 2010). Aside from motor speech symptoms (dysarthria) – which as a rule occur after remission of mutism – a variety of concomitant non-motor language disturbances have been identified. These include word-finding difficulties (Levisohn et al., 2000; Aarsen et al., 2004), agrammatism (Siffert et al., 2000; Riva and Giorgi, 2000), disrupted language dynamics (Siffert et al., 2000; Ozimek et al., 2004), comprehension deficits (Levisohn et al., 2000; Cornwell et al., 2003), reading (Scott et al., 2001) and writing problems (Aarsen et al., 2004). At the emotional and affective level, frequent mention is made of adynamia and symptoms indicating inhibition of frontal lobe functioning such as a lack of initiative, aspontaneity, apathy (Pollack, 1997), disinterest (Steinlin et al., 2003), emotional unsteadiness (Catsman-Berrevoets et al., 2003), flattened affect, inadequate emotional coping (Ozimek et al., 2004), diminished eye contact, and withdrawal (Daniels et al., 2005). In marked contrast to the view that the PFS is a transient phenomenon (Van Dongen et al., 1994), long-term cognitive consequences of the PFS that have been identified include scholarly underachievement and major cognitive sequelae such as a significant decline of general intelligence, executive dysfunction, disrupted memory, attentional deficits, and distorted spatial cognition (Levisohn et al., 2000; Steinlin et al., 2003; De Smet et al., 2009).

Several risk factors for the development of the PFS have been identified such as the type and size of the lesion (Catsman-Berrevoets et al., 1999), length of the vermian incision (Dailey et al., 1995), midline tumour location (Pollack et al., 1995), tumour location adjacent to the fourth ventricle and post-surgical oedema of the pontine tegmentum (Van Dongen et al., 1994), and the occurrence of postoperative hydrocephalus or meningitis (Humphreys, 1989; Ferrante et al., 1990; Salvati et al., 1996). Despite the growing interest in PFS, its pathophysiological substrate remains to be clarified. Several hypotheses have been put forward to explain PFS: 1) postoperative vasospasms of the arteries supplying the cerebellum and the brainstem leading to ischaemia and subsequent oedema (Ferrante et al., 1990; Ildan et al., 2002; Maffei et al., 2005), 2) transient dysfunction of the A9 and A10 mesencephalic dopaminergic cell groups and ascending activating reticular system (Catsman-Berrevoets et al., 1992) and, 3) bilateral surgical damage to the dentate and interpositus nuclei (Pollack et al., 1995; Kusano et al., 2006) or to the afferent and/or efferent pathways passing through these nuclei (Crutchfield et al., 1994; Ersahin et al., 1996). The hypothesis that the cognitive and behavioural dysfunctions associated with the PFS may result – as reflected by the phenomenon of cerebello-cerebral diaschisis – from a temporarily functional depression of the reciprocal pathways that connect the cerebellum with the limbic circuitry and the prefrontal, temporal and parietal association cortices is attracting growing attention (Fig. 1) (Germano et al., 1998; Baillieux et al., 2006; Mariën et al., 2001, 2003, 2008, 2009, 2010; Morris et al., 2009; Catsman-Berrevoets and Aarsen, 2010; Miller et al., 2010).

In sharp contrast to children, PFS has only very occasionally been documented in adults. This paper intends to contribute to the limited amount of information that is available. Its objective is to describe the neurobehavioural and neuroradiological findings during a 11.5-months follow-up period in a 38-year-old female patient who presented with cognitive, behavioural and affective symptoms before and after resection of an ependymoma in the posterior fossa. To the best of our knowledge, this is the very first study in which an adult patient was investigated by means of pre- and postoperative single photon emission computed tomography (SPECT) and longitudinal neuropsychological follow-up investigations.

2. Case report

2.1. History

2.1.1. Preoperative findings

A 38-year-old right-handed native Dutch-speaking woman was referred to the department of neurology because she had been suffering from slightly progressive symptoms for at least seven months. These consisted of fatigue and paroxysms of vertigo, nausea, vomiting and headaches. As noted by her husband, she also presented with behavioural and affective changes, mainly characterised by decreased motivation, unconcern, social withdrawal, and incidences of verbal aggression and inappropriate verbal remarks. The clinical neurological examination on admission revealed an alert and cooperative patient who was not concerned about her medical condition and who displayed mild verbal disinhibition (use of foul language, inappropriate verbal reactions, cursing). Sensory examination was

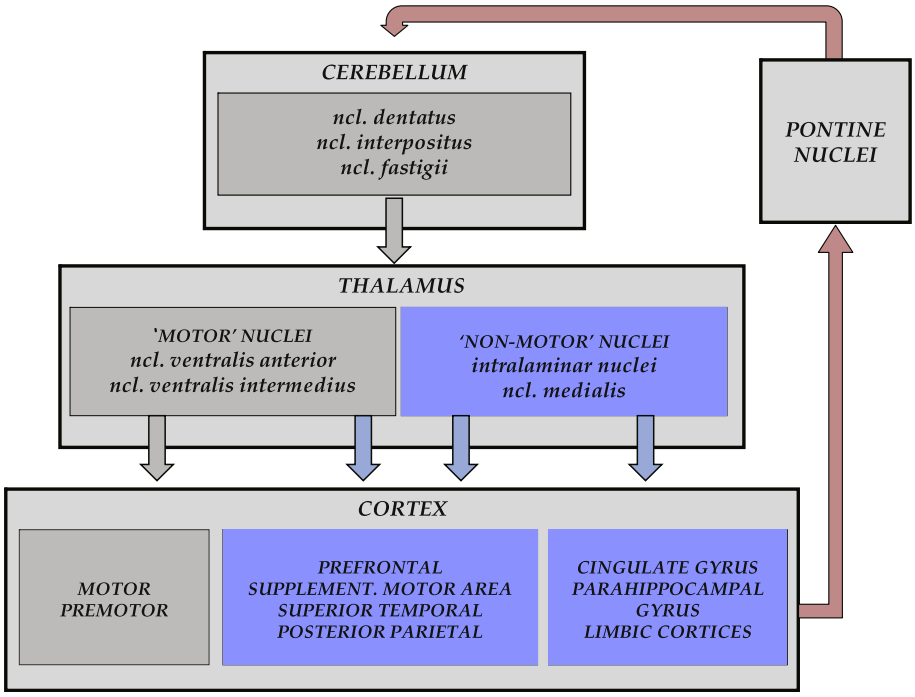

Fig. 1 – Diagram depicting cerebello-cerebral connectivity network subserving cognitive and affective processes. The feedbackward or efferent loop originates from the deep nuclei of the cerebellum which project to the motor (grey arrows) and non-motor (blue arrows) nuclei of the thalamus. In turn, the motor nuclei of the thalamus (ncl. ventralis anterior and intermedius) project to motor and premotor cortices (grey arrows) but also to non-motor areas among which the prefrontal cortex, the supplementary motor area, the superior temporal and posterior parietal regions (blue arrows). The non-motor nuclei of the thalamus project to the cingulate gyrus, the parahippocampal region and the limbic cortices (blue arrows). The feedforward or afferent system of the cerebello-cerebral circuit is composed of corticopontine and pontocerebellar mossy fibre pathways (red arrows) (after Schmahmann and Pandya, 1997).

unremarkable. Examination of coordination by finger-to-nose and heel-to-knee tests was normal but slight gait ataxia and unsteadiness were noticed. Eye movements were normal and cranial nerves were intact. Tendon reflexes were normal. Plantar response was flexor bilaterally. Speech was well-articulated and neither dysarthric nor apraxic. Auditory-verbal comprehension, repetition and naming were within normal limits.

Growth and developmental milestones were normal. The patient had an educational level of 12 years and worked as a clerk in a supermarket. Medical history consisted of an ovarium carcinoma for which she was operated and treated with radiotherapy at the age of 12 years. A hysterectomy was carried out at the age of 29 years.

Cranial computed tomography (CT) scan findings on admission were confirmed by an magnetic resonance imaging (MRI) scan of the brain, which revealed a large inhomogeneous and hypo-intense mass lesion with a diameter of approximately 27.5 mm in the fourth ventricle (Fig. 2A–F). A secondary hydrocephalus of the third and lateral ventricles was found as well. No clear signs of periventricular oedema were found (Fig. 3A–F). There was no evidence of supratentorial damage.

Quantified Technetium-99m-Ethyl Cysteinate Dimer (Tc-99m-ECD) SPECT scan of the brain was carried out two days before surgery. Using a previously fixed butterfly needle 740 MBq (20 mCi) Tc-99m-ECD was administered to the patient sitting in a quiet and dimmed room, eyes open and ears unplugged. Acquisition was started 40 min after injection using a three-headed rotating gamma camera system (Triad 88; Trionix Research Laboratory, Twinsburg, Ohio, USA) equipped with lead super-fine fanbeam collimators with a system resolution of 7.3 mm FWHM (rotating radius 13 cm). Projection data were accumulated in a 128 × 64 matrix, pixel size 3.56 mm, 15 sec per angle, 120 angles for each detector (3° steps, 360° rotation). Projection images were rebinned to parallel data, smoothed and reconstructed in a 64 × 64 matrix, using a Butterworth filter with a high cut frequency of .7 cycles/cm and a roll-off of 5. No attenuation or scatter correction was carried out. Trans-axial images with a pixel size of 3.56 mm were anatomically standardised using statistical parametric mapping (SPM) and compared to a standard normal and standard deviation (SD) image obtained from ECD perfusion studies in a group of 15 normally educated healthy adults consisting of 8 men and 7 women with an age ranging from 45 to 70 years. Using a 31 region-of-interest (ROI) template, Z-scores (SD) were calculated for each region. A regional Z-score of >2.0 was considered significant. In comparison to normal database findings quantified Tc-99m-ECD SPECT results showed decreased perfusion in the vermis (−2.91) and the left cerebellar hemisphere (−1.89 SD) as well as an area of hypoperfusion bilaterally in the cingular gyrus (left = −4.50; right = −4.30), the

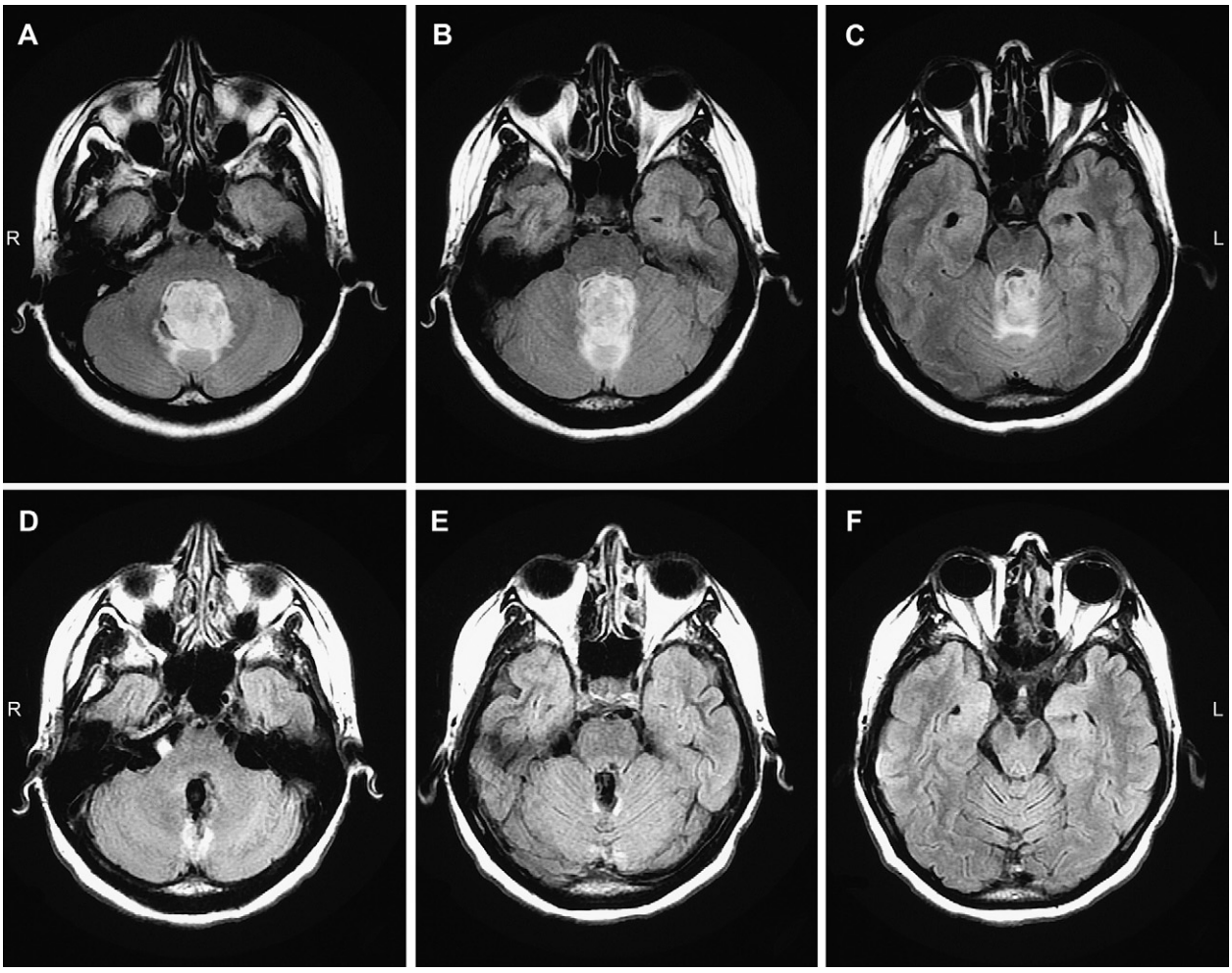

Fig. 2 – Brain MRI axial T2-weighted FLAIR slices on admission (A–C) disclosing a hyperintense tumoural mass lesion in the fourth ventricle with a diameter of approximately 27.5 mm and early postoperative T2-weighted FLAIR slices (D–F) showing near-total resection of a low-grade ependymoma.

thalamus (left = −2.81; right = −2.49), and both caudate nuclei (left = −3.72; right = −3.76) (Table 1). Postoperative Tc-99m-ECD SPECT studies were performed 6 days, 1.5, 3.5, 7.5 and 9.5 months after surgery.

Formal neuropsychological assessments were performed 2 days before surgery and during 11.5 months of postoperative follow-up. Cognitive results are discussed in detail under a separate heading below.

Near complete resection of the tumour was achieved after placement of an external ventricular drain and incision from the inion to C2–C3 with an incision of the vermis of 3 cm. Anatomopathological examination of the tumour specimen disclosed a low-grade (grade II) ependymoma.

2.1.2. *Postoperative findings*

The immediate postoperative course was uneventful. The patient remained sedated and intubated for the next two days after the operation. On the third day after surgery, the clinical neurological examination revealed an alert and cooperative patient with severe ataxic dysarthria characterised by vowel and consonant distortions, irregularly articulatory breakdowns, scanning of speech, prominent bradylalia and harsh voice quality. Although verbal output was severely distorted at the articulatory level, the patient was able to speak spontaneously with syntactically correct phrases and sentences. Bedside visual confrontational naming of five objects (e.g., pencil, stethoscope) and five colours was correct. Auditory-verbal comprehension was intact for daily language use. Three step commands were completed flawlessly. Spatial and temporal orientation was unimpaired and the recall of three auditorily presented words was intact. A peripheral right facial nerve paresis and a mild right brachial paresis (4/5) were found. The plantar response was flexor bilaterally. Midline fixation of both eyes was present when the head was rotated in all directions. She could not visually follow objects moving in front of her and she complained of diplopia. A divergent strabismus was found. Finger-to-nose and heel-to-knee tests were bilaterally disturbed and dysdiadochokinesia and intentional tremor were found. Gait and equilibrium were not examined at that time. Sensory examination was normal. The patient sporadically experienced complex visual hallucinations that mainly consisted of colourful cats running around

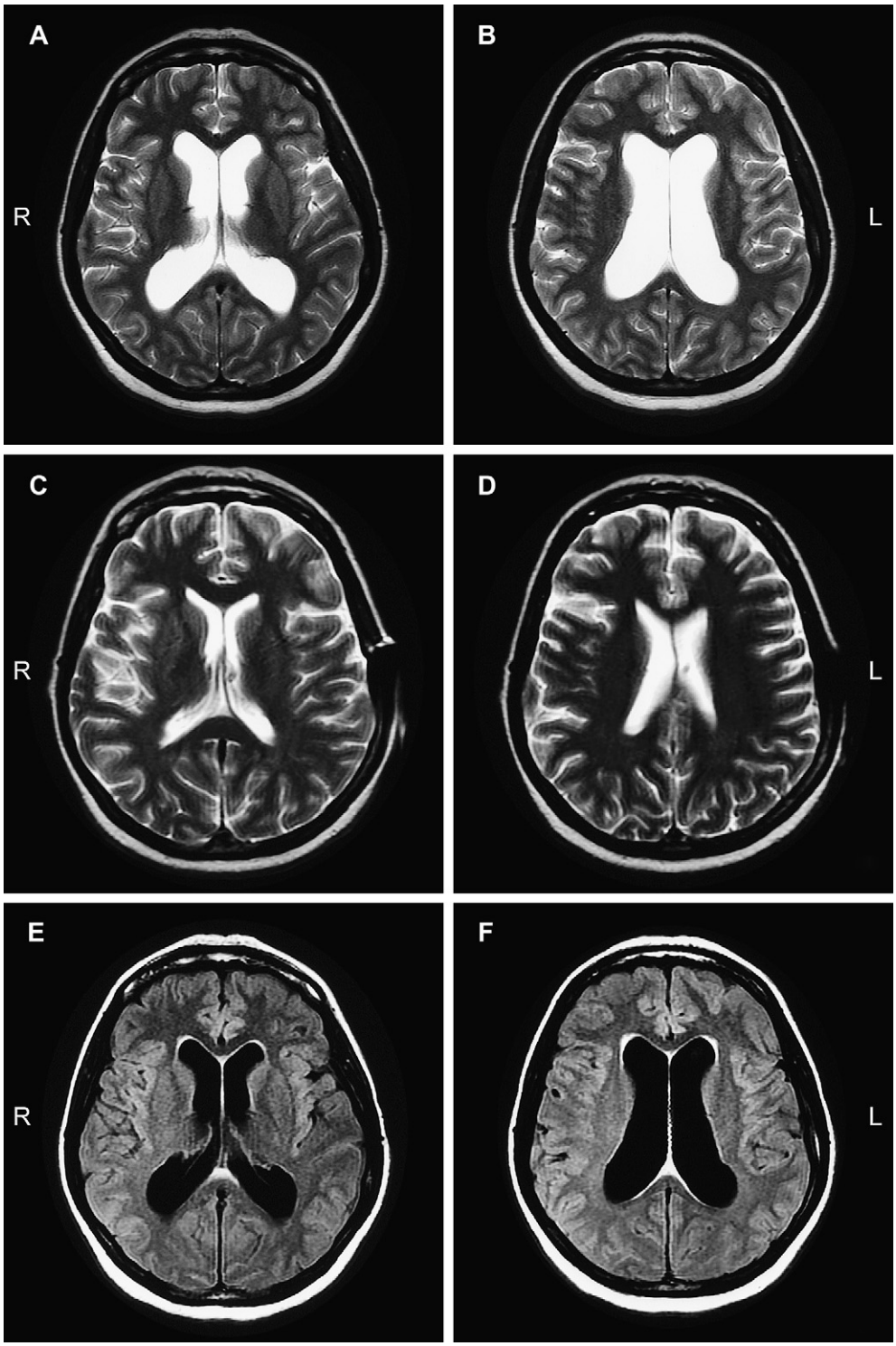

Fig. 3 – MRI axial T2-weighted supratentorial images disclosing signs of hydrocephalus before the operation (A–B) and normalisation of the ventricular volumes in the early postoperative phase after shunting (C–D). Preoperative MRI axial T2-weighted supratentorial FLAIR slices did not disclose periventricular high signal lesions, excluding important intracranial hypertension in the presence of hydrocephalus (E–F).

and of faces of unknown people staring at her through the windows of the hospital room. Mood and affect were not appropriate. She remained unconcerned about her medical condition and acted in a rather euphoric way. She often responded to external stimuli in a frontal-like, disinhibited manner and presented with inappropriate behaviour, over-familiarity or flamboyant and impulsive actions. Physical therapy and intensive speech therapy were started. Repeat quantified Tc-99m-ECD SPECT scan of the brain performed six days after tumour surgery again showed decreased perfusion

Table 1 – Pre- and postoperative quantified Tc-99m-ECD SPECT brain perfusion study results expressed in ≥−1.5 SDs during 9.5 months of follow-up.

	2 days preoperative		6 days postoperative		41 days postoperative		3.5 months postoperative		7.5 months postoperative		9.5 months postoperative	
	L	R	L	R	L	R	L	R	L	R	L	R
Prefrontal lateral					−1.89		−2.96	−2.34	−2.02	−2.91	−1.92	−2.00
Prefrontal medial					−2.18	−2.35	−1.96	−2.92	−2.53	−3.42		−2.28
Frontal inferior lateral		−1.52			−1.56	−2.12			−2.05	−2.30	−1.73	−2.08
Frontal inferior medial		−1.79				−1.60						−1.50
Cingulate gyrus	−4.50	−4.30	−3.58	−3.43		−1.72			−1.67	−1.87		
Thalamus	−2.81	−2.49	−1.70									
Caudate nucleus	−3.72	−3.76	−3.53	−2.79								
Occipital			−1.97									
Cerebellum	−1.89											
Vermis	−2.91		−3.42		−3.40		−2.71		−3.09		−3.05	

L = left; R = right.

in the vermis (−3.42), both cingulate gyri (left = −3.58; right = −3.43) and both caudate nuclei (left = −3.53; right = −2.79). The perfusional deficits in the thalami had resolved (<−2SD). A relative hypoperfusion in the left occipital lobe almost reached significance (−1.97) (Table 1).

During the next days the frequency of the hallucinations increased and the patient progressively developed other psychotic and confusional symptoms consisting of agitation, anxiety, restlessness, pseudo-bulbar-like crying and laughter, shouting behaviour, withdrawal, disrupted and diminished contact, emotional lability, apathy, behavioural regression, childish and disinhibited actions. An MRI of the brain performed 12 days after surgery did not disclose any new findings. Apart from a decrease of the ventricular volumes, near-total resection of the tumoural mass in the fourth ventricle and a collection of blood in the surgical area were observed (Fig. 2D–F). No supratentorial lesions were found. The patient did not receive any analgesic drug that may induce hallucinations. No meningeal signs or fever were found. In addition, laboratory test results did not disclose any evidence for an infection or electrolyte imbalance that might explain the delirant state. A treatment with risperidone (2 × .5 mg daily) was started 13 days after surgery but discontinued within one week because of a further decrease of contact and an increased apathy. Interruption of the treatment did, however, not result in a clinical improvement. Because of an elevation of intraventricular pressure (18 cm H_2O), additionally demonstrated on CT by an increase of the ventricular volumes, the external drain, which was inserted via the right frontal region, was removed 24 days after tumour resection and a permanent ventriculoperitoneal shunt was installed. Although several repeat CT scans and an MRI of the brain showed a normalisation of the ventricular volumes (Fig. 3), confusional symptoms and aberrant frontal-like behavioural characteristics progressively worsened over the next days. The patient, however, did not go through any infectious event and no evidence was found for iatrogenic meningitis. Repeat cerebrospinal fluid and blood examinations showed no evidence of infection. Hardly any contact could be established and the patient only responded after strong incentives with one or two word phrases. Thirty-one days after tumour surgery, the patient evolved to a state of complete mutism. As the loss of speech was accompanied by a substantial decrease of volitional motor activity, lethargy, moderately preserved vigilance functions, unimpaired visual tracking of external stimuli and general apathy, the condition matched a diagnosis of akinetic mutism. Again no clinical nor laboratory evidence was found to explain this evolution. Repeat CT scan of the brain did not disclose any new findings. Repeat quantified Tc-99m-ECD SPECT of the brain 41 days post-surgery showed a remission of the perfusional deficits at the subcortical level (caudate nuclei). However, in contrast to prior findings the perfusional changes grossly affected the frontal brain regions. A significant hypoperfusion was observed bilaterally in the medial prefrontal areas (left = −2.18; right = −2.35) and the right lateral inferior frontal region (−2.12). The perfusional defect at the vermis remained unchanged (−3.40) (Table 1).

After 12 days of akinetic mutism the clinical condition started to change. During the next month, akinetic mutism alternated with periods in which verbal responses could be elicited. However, during these intervals the patient mostly did not generate any speech spontaneously. Her movements were slowly executed and the speed of cognitive processing was marked by bradyphrenia. Mitigated echo-answers, contaminated by dysarthria, and adequate (non-)verbal reactions to simple verbal commands could be evoked only after stimulation. At the affective and behavioural level, the patient often responded with drama and outbursts of intense and uncontrollable pseudo-bulbar-like crying and laughter. During these attacks contact was difficult to maintain. After one month of fluctuations a treatment with a non-ergoline dopamine-agonist (ropinirole hydrochloride; 3 × .25 mg daily increased over 6 weeks to 3 × 2 mg) was started which rapidly established normalisation of responsiveness, remission of apathy and marked improvement of distorted language dynamics and mental slowness. The patient, although still severely dysarthric, spontaneously used short phrases and started showing some interest again in her immediate environment. Nevertheless, she was still very

unstable with regard to behavioural, emotional and affective regulation. She acted in a very childish and theatrical way and presented with inappropriate, disinhibited actions. Outbursts of pseudo-bulbar-like crying and laughing re-appeared.

Five weeks after medical treatment and intensive cognitive therapy, a subset of neuropsychological tests was carried out with results showing significant dysfunctions in multiple cognitive domains. Repeat quantified Tc-99m-ECD SPECT of the brain during this phase (3.5 months post-surgery) revealed significant perfusion deficits bilaterally distributed in the lateral prefrontal (left = −2.96; right = −2.34) and medial prefrontal (left = −1.96; right = −2.92) brain regions and the vermis (−2.71) (Table 1). However, at a daily dose of 6 mg ropinirole hydrochloride the patient again developed psychotic symptoms (agitation, confusion, paranoia, visual hallucinations, delusions, and verbal and physical aggression). Progressive lowering of the daily dose to .75 mg with discontinuation of the drug after 11 weeks of treatment gradually improved the psychotic symptoms but caused the frontal-like behavioural and affective disturbances that had persisted during follow-up to re-appear.

Repeat quantified Tc-99m-ECD brain SPECT results at 7.5 and 9.5 months post-surgery remained basically unchanged, showing persistent perfusion deficits in the frontal lobes and the vermis. Repeat MRI of the brain at 6 and 9 months post-surgery did not disclose any new findings. At the motor level, the patient's neurological condition improved but distinct midline cerebellar dysfunction, presenting as truncal ataxia and gait ataxia, resulted in tandem gait which made independent walking impossible. Appendicular ataxia was also demonstrated by ataxic and dysmetric finger-to-nose and heel-to-shin tests and the presence of dysdiadochokinesis. Cerebellar tremor was also present. The peripheral right facial nerve paresis and diplopia with divergent strabismus persisted as well. One year after tumour surgery the patient was referred to a neurological rehabilitation centre without any significant improvement. Three years after surgery, the patient is able to walk with a rollator. Mood swings and psychotic episodes with paranoia persist. The clinical neurological examination remains unchanged. Repeat MRI of the brain shows no brainstem lesion and no tumour recidive.

2.2. *Neurocognitive investigations*

Formal neurocognitive investigations were carried out one day prior to surgical resection of the tumour and repeated 3.5, 5, 7 and 11.5 months after the operation (Table 2). The neuropsychological test battery consisted of standardised tests including: the Mini Mental State Examination (MMSE) (Folstein et al., 1975), the Wechsler Adult Intelligence Scale-III (WAIS-III) (Wechsler, 1997), the Wechsler Memory Scale-Revised (WMS-R) (Wechsler, 1987), the Progressive Matrices (RPM) (Raven, 1976), the Repeatable Battery for the Assessment of Neuropsychological Status (RBANS) (Randolph, 1998), the Hierarchic Dementia Scale (HDS) (Cole and Dastoor, 1987), the Stroop Colour-Word test (Golden, 1978; Lezak, 1995), the Wisconsin Card Sorting Test (WCST) (Heaton et al., 1993), the D2 test of sustained visuo-motor attention (Brickenkamp and Zillmer, 1998), and the Trail Making Test (Reitan, 1958). Handedness was formally assessed by means of the Edinburgh Inventory (Oldfield, 1971). The neurolinguistic test battery consisted of subtests of the Boston Diagnostic Aphasia Examination (BDAE) (Goodglass and Kaplan, 1983), the Boston Naming Test (BNT) (Mariën et al., 1998) and a semantic verbal fluency task (1 min generation of names of animals, means of transport, vegetables, clothes) (unpublished norms). Video-taped samples of spontaneous and conversational speech were collected for later analysis.

2.2.1. *Preoperative findings*

Formal assessment of handedness by means of the Edinburgh Inventory (Oldfield, 1971) revealed a strong and consistent right hand preference, reflected by a laterality quotient of +100. General cognitive screening was normal as witnessed by full marks on the MMSE. Visuospatial problem-solving, assessed by means of the RPM revealed a normal non-verbal IQ level of 89. The WMS-R profile showed a slight discrepancy of 15 points between the verbal (=109) and visual immediate memory index (=94) which were both normal. A delayed memory index of 74 (=−1.7 SD) indicated depression of recent memory. Attentional skills were disrupted (WMS-R concentration index = 69; −2.1 SD). Auditory as well as visual attention-span scored within the defective range (<percentile 5). Sustained visuo-motor attention was also decreased. On the D2 letter cancellation task a low average score of percentile 11 was found. As demonstrated by the WCST, frontal planning and conceptual organisation of goal-oriented cognitive strategies was impaired. The ability to inhibit a competing and more automatic response set was intact (Stroop Colour-Word). Visual search and sequencing (Trail Making Test) were also normal. At the linguistic level, auditory comprehension was normal (BDAE commands = 15/15). Visual confrontation naming was disrupted (BNT = 49/60; −2.1 SD). The majority of naming errors were of mnestic origin (6 out of 11 errors). The semantic word fluency task attracted a normal score. No reading errors were made in an oral sentence reading task (BDAE = 10/10). The mechanics of writing and spelling to dictation were also normal (BDAE = 10/10).

2.2.2. *Postoperative findings*

Formal cognitive investigations could not be reliably carried out during the first three and a half months after the operation. Concise cognitive testing 3.5 months postoperatively revealed a general cognitive decline reflected by a range of impairments in various cognitive domains (Table 2). The MMSE was severely deficient (20/30; −4.4 SD). The HDS revealed significant deficits in the following domains: praxis (ideomotor, constructional, drawing apraxia), gnosis (visual object agnosia), language (comprehension, naming, reading, writing), memory (registration, recent memory), orientation and calculation.

At 5 months post-surgery no significant change was found (Table 2). A speech sample consisting of 8 min of conversational speech was video-taped for further analysis. As demonstrated in Table 3 and Fig. 4, conversational skills were severely disturbed.

Table 2 – Neurocognitive test results.

Neurocognitive tests	Preop.	3.5 m postop.	5 m postop.	7 m postop.	11.5 m postop.	Mean (SD)
	Score (SD)	Score (SD)	Score (SD)	Score (SD)	Score (SD)	
Mini Mental State Examination	30	20 (−4.4)	20 (−4.4)	21 (−3.9)	23 (−2.8)	28 (1.8)
Intelligence						
WAIS-III						
Verbal IQ				59 (−2.7)	71 (−1.9)	100 (15)
Vocabulary				2 (−2.6)	5 (1.6)	10 (3)
Similarities				2 (−2.6)	4 (−2.0)	10 (3)
Arithmetics				4 (−2.0)	4 (−2.0)	10 (3)
Digit span				4 (−2.0)	6 (−1.3)	10 (3)
Information				5 (−1.6)	7 (−1.0)	10 (3)
Comprehension				3 (−2.3)	4 (−2.0)	10 (3)
Performal IQ				56 (−2.9)	53 (−3.1)	100 (15)
Picture completion				4 (−2.0)	4 (−2.0)	10 (3)
Block design				3 (−2.3)	1 (−3.0)	10 (3)
Matrix reasoning				3 (−2.3)	4 (−2.0)	10 (3)
Picture arrangement				2 (−2.6)	2 (−2.6)	10 (3)
Full Scale IQ				59 (−2.7)	63 (−2.5)	100 (15)
Progressive Matrices	89			75 (−1.6)	79 (−1.4)	100 (15)
Memory						
WMS-R						
Visual Memory Index	94				81 (−1.3)	100 (15)
Verbal Memory Index	109				85 (−1.0)	100 (15)
Global Memory Index	103				82 (−1.2)	100 (15)
Delayed Recall Index	74 (−1.7)				105	100 (15)
RBANS						
Immediate memory				69 (−2.0)	97	100 (15)
Visuoperceptual cognition				58 (−2.8)	60 (−2.7)	100 (15)
Language				74 (−1.7)	54 (−3.1)	100 (15)
Attention				40 (−4.0)	43 (−3.8)	100 (15)
Delayed Memory				44 (−2.4)	83 (−1.1)	100 (15)
Hierarchic Dementia Scale						
Orienting		10	10			10 (0)
Prefrontal		10	10			10 (0)
Ideomotor		8 (−8.4)	8 (−8.4)			9.94 (.23)
Looking		6 (>2)	10			10 (0)
Ideational		10	7 (−17.5)			9.97 (.17)
Denomination		9 (−5.7)	9 (−5.7)			9.97 (.17)
Comprehension		4 (>2)	9 (−5.7)			9.97 (.17)
Registration		6 (−11)	8 (−5.3)			9.86 (.35)
Gnosis		9 (−2.5)	9 (−2.5)			9.92 (.37)
Reading		6 (>2)	10			10 (0)
Orientation		6 (>2)	10			10 (0)
Construction		8 (>2)	4 (>2)			10 (0)
Concentration		3 (−12.8)	5 (−9.0)			9.69 (.52)
Calculation		2 (−34.5)	7 (−12.8)			9.94 (.23)
Drawing		0 (−18.9)	3 (−13.1)			9.81 (.52)
Motor		10	10			9.58 (1.05)
Remote memory		10	6 (>2)			10 (0)
Writing		0 (−7.9)	0 (−7.9)			9.94 (1.26)
Similarities		10	10			9.72 (.7)
Recent memory		6 (−4.0)	6 (−4.0)			9.5 (.88)
Concentration						
WMS-R concentration	69 (−2.1)					
D2 test (pct)	Pct 11					
Language						
Boston Naming Test	49 (−2.1)		48 (−2.5)	48 (−2.5)	48 (−2.5)	54.5 (2.59)
Verbal fluency						
Total Semantic Generation	70			33 (−2.0)	34 (−1.9)	59.7 (13.27)
Animals. 1 min	20			7	9	
Transportation. 1 min	14			7	6	
Vegetables. 1 min	16			6	6	

(*continued on next page*)

Table 2 – (*continued*)

Neurocognitive tests	Preop.	3.5 m postop.	5 m postop.	7 m postop.	11.5 m postop.	Mean (SD)
	Score (SD)	Score (SD)	Score (SD)	Score (SD)	Score (SD)	
Clothing. 1 min	20			12	11	
Perseverations	4			1	1	
Executive Functioning						
Wisconsin Card Sorting	0 cat.				0 cat.	
Stroop Colour-Word Test						
Card I	80			/	<1	50
Card II	50			/	<1	50
Card III	40			/	<1	
Trail Making						
A	50			Pct < 10	<10	50
B	30			/	<10	50

Preop. = preoperative; Postop. = Postoperative; m = months; Pct = percentile; N = number; WAIS-III = Wechsler Adult Intelligence Scale 3rd Edition; VIQ = verbal intelligence quotient; PIQ = performance intelligence quotient; WMS-R = Wechsler Memory Scale-Revised; RBANS = Repeatable Battery for the Assessment of Neuropsychological Status; HDS = Hierarchic Dementia Scale; cat. = correct categories.

Conversational turn taking was marked by echolalic utterances (46.2%). A total of 13.2% of her utterances were palilalic in nature. Only 40.6% of all utterances could be considered as new input. Another extract of conversational speech consisting of 300 words was analysed to calculate mean length of utterance (MLU) and type token ratio (TTR). In comparison to a normal MLU range of 5.7–10.7 (Boxum and Zwaga, 2007), a reduced MLU of 4.7 was found. TTR of nouns was normal (.8; mean .75, range .54–.86), whereas TTR of verbs (.6) was slightly below the normal range (mean .74, range .63–.86). Motor speech analysis by means of the dysarthria protocol developed by Darley et al. (1975), revealed a slow speaking rate, prolonged phonemes and intervals, imprecise articulation of consonants and consonant clusters, low pitch, monopitch, strained-strangled voice and excess and equal stress. Furthermore, speech tempo was investigated and following Laver (2002) and Verhoeven et al. (2004), a distinction was made between speaking rate and articulation rate. Speaking rate represents the number of syllables per second (syll/s) including filled and silent pauses, whereas in articulation rate these silent pauses are excluded. Based on an interview of 20 min, the patient's average articulation rate was found to be 2.12 syll/s and speaking rate was 1.82 syll/s. These values are well below the articulation and speaking rates of the average female speakers in Belgium which amount to 4.50 syll/s and 4.01 syll/s respectively (Verhoeven et al., 2004).

Table 3 – Analysis of an 8 min sample of conversational speech 5 months after surgery, containing 195 words.

	Number of utterances	Number of words	Proportion
Echolalic utterances	19	91	46.2%
Palilalic utterances	7	26	13.2%
New utterances	20	80	40.6%
Total	46	197	MLU = 4.3

MLU = mean length of utterance.

Seven months post-surgery more extensive neurocognitive examinations could be carried out. The WAIS-III revealed a pathological full scale IQ of 59 (−2.7 SD) (Table 2). At the verbal level, a defective VIQ of 59 (−2.7 SD) was found. The performance IQ was 56 (−2.9 SD). Scaled scores of −1.5 or below −1.5 SD were obtained for all verbal and performance subtests. In agreement with these findings, the RBANS showed multiple cognitive domain impairments. In comparison with the preoperative results, BNT results did not significantly change. Results on the semantic verbal fluency task (33 items) significantly decreased (>−2 SD). A score below percentile 1 on the

I: Madam?
P: Madam…
I: You know that we visit you regularly and that we use a video-camera to record our conversations.
P: Video-camera…to record…our conversations.
I: The purpose is to do that today too.
P: The purpose is to do that today too.
I: Do you agree with it?
P: Do you agree with it… Yes.
I: Okay, that's a good start.
P: Oh yes.
I: Did Bert visit you yesterday?
P: Yesterday, yesterday…
I: Did you go home for the weekend?
P: Yes
I: Tell me, how was the weekend?
P: A very nice weekend…
I: Great. What did you do?
P: We collected Easter eggs…
We saw the Easter bells.
I: Saw them?
P: Saw them.
I: Where did you see them?
P: We did see them, we did see them, did see them…
I: Did you come back here on Sunday?
P: Sunday I did come back here.
I: So, then Bert dropped you off.
P: then Bert dropped me off.
I: And then both of you went to the seventh floor?
P: then both of us went to the seventh floor.

I: interlocutor, P: patient

Fig. 4 – Literal translation of 2 min of conversational speech extracted from an 8 min sample obtained 5 months after posterior fossa tumour surgery.

Stroop Colour-Word test indicated disrupted ability to inhibit a competing and more automatic response set.

At 11.5 months post-surgery a mild improvement of verbal functions and memory was found (Table 2). The VIQ increased with 12 points to a level of 79 (−1.9 SD) but the PIQ did not improve (=53). Mnestic functions improved but WMS-R immediate memory levels remained slightly depressed. Visuo-spatial/constructional skills, language and attention did not substantially improve (RBANS) nor did naming (BNT), verbal fluency and frontal problem-solving (WCST).

3. Discussion

To the best of our knowledge, this study for the first time reports the pre- and postoperative clinical, neurocognitive and SPECT findings in an adult patient who following surgical resection of an ependymoma in the posterior fossa developed symptoms consistent with a diagnosis of PFS. In sharp contrast to the children in which the incidence of the PFS after posterior fossa tumour resection is estimated to range between 8% (Pollack et al., 1995; Dailey et al., 1995; Van Calenbergh et al., 1995) and 31% (Catsman-Berrevoets et al., 1999, 2003; Van Mourik et al., 1998; Robertson et al., 2006; Küpeli et al., 2011), the condition has only very occasionally been documented in adults. A meticulous survey of the literature on cerebellar mutism in adults – patients of 18-years-old and older according to the WHO criteria – following posterior fossa surgery yielded reports on only 21 adult patients, published in the period 1969 to January 2011. This is only one tenth of the total number of children with PFS. The characteristics of these adult cases are summarised in Appendix 1 [case reference numbers 1 to 21].

Given the rarity of the PFS in the adult patient population and the many unsolved questions relating to its semiology and underlying pathophysiological substrate, several observations in our patient deserve some further discussion: 1) the preoperative neurobehavioural findings, 2) the postoperative progression to full-blown PFS, 3) the "relapsing-remitting" course of akinetic mutism resolving after treatment with a non-ergoline dopamine-agonist, 4) the phenomenon of pathological laughing and crying (PLC), 5) the postoperative cognitive decline and persisting CCAS, 6) pre- and post-operative hydrocephalus as a possible risk factor for PFS, and 7) involvement of the cerebello-cerebral network as evidenced by quantified ECD SPECT. Finally, a comparison between PFS in children and adults is made.

3.1. *Preoperative findings*

Preoperative neurocognitive and behavioural investigations have not been systematically carried out in the adult (see Appendix) or in the paediatric patient population with posterior fossa tumours. However, formal cognitive assessments and behavioural observations in this patient revealed a range of clinically significant cognitive, affective and behavioural symptoms consistent with CCAS. It might be hypothesised that preoperative CCAS reflects tumour induced disruption of the cerebello-cerebral network, possibly implying a higher risk for the development of the PFS.

3.2. *Postoperative findings: the development of akinetic mutism*

In the immediate postoperative phase the patient presented brainstem and cerebellar symptoms associated with a dysregulation of affect, disinhibited behaviour, hypersomnolence and episodes of vivid complex visual hallucinations. Although a broad range of neurobehavioural abnormalities may occur after posterior fossa tumour surgery (Baillieux et al., 2006), peduncular hallucinations, a term coined by Van Bogaert (1927), have not been described before in the adult population and have only been exceptionally observed in children. Indeed, only Kumar and Kaur (2000) reported a brief period of peduncular hallucinations after midline posterior fossa medulloblastoma resection in a four-year-old boy. The pathophysiological substrate of this phenomenon remains to be clarified, but the frequent combination with oculomotor disturbances, dysarthria, confusion, hypersomnolence and ataxia (Benke, 2006) strongly indicates midbrain dysfunction. However, repeat MRI in this patient did not disclose any damage in the midbrain, the pons or the diencephalon and this is similar to the findings of Kumar and Kaur (2000). As a result, the association of peduncular hallucinations and brainstem symptoms might reflect functional disruption of: 1) the structurally intact brainstem due to traction trauma during tumour resection (Kumar and Kaur, 2000) or 2) the cerebellar-mesencephalic network following a reduced input of excitatory impulses from the surgically damaged cerebellum.

No medical or pharmacological reasons were found for the development of confusional symptoms, psychotic behaviour and lethargy. The patient was not treated with analgesic drugs that may induce hallucinations and laboratory test results did not disclose any evidence for an infection or electrolyte imbalance. Thirty-one days after the operation "akinetic mutism" arose which is quite exceptionally recorded in the context of PFS. To the best of our knowledge, only two paediatric cases (Caner et al., 1999; Adachi et al., 2005) and one adult patient (Moore, 1969) have been reported with akinetic mutism following posterior fossa surgery.

3.3. *Relapsing-remitting akinetic mutism*

In this patient akinetic mutism temporarily resolved and reappeared after short episodes of significantly improved but still diminished contact. These 'pseudo-lucid intervals' lasted for several hours to maximum one day. To the best of our knowledge a relapsing-remitting course of akinetic mustism has not been reported before.

Since dopamine-agonists have been proven useful to treat akinetic mutism of variable aetiology (Catsman-Berrevoets and van Harskamp, 1988; Echiverri et al., 1988; Mateo-Sierra et al., 2005; Caner et al., 1999; Combarros et al., 2000; Alexander, 2001; Kim et al., 2007; Spiegel et al., 2008), a treatment with ropinirole hydrochloride was started one month after "relapsing-remitting akinetic mutism" occurred. This non-ergoline dopamine-agonist that specifically binds with D2 and D3 receptors induced a complete remission of akinetic mutism and significantly ameliorated

the patient's decreased level of responsiveness and alertness. Although an improvement of mental speed, language dynamics, behaviour and affect was found as well, CCAS persisted. The favourable response to treatment with a non-ergoline dopamine-agonist seems to corroborate the view that a decrease of dopaminergic input to the prefrontal cortex, as a result of cortical or subcortical lesions affecting the mesocorticolimbic dopaminergic projections, might be an important mechanism involved in akinetic mutism (Cummings, 1993).

3.4. *The cerebellum and PLC*

Postoperatively the patient presented frequent outbursts of pseudo-bulbar-like crying and laughter. These uncontrollable emotional disruptions occurred in the absence of any apparent triggering stimuli and can therefore be considered to represent PLC (Parvizi et al., 2001; Wild et al., 2003). Wilson (1924) hypothesised that PLC is caused by a loss of voluntary inhibition of a presumed centre of laughing located in the upper brainstem. This view has been substantially challenged by neuropathological findings and evidence about the neurobiological basis of emotions and feelings. Parvizi et al. (2001) described a patient who presented PLC as a primary result of a right cerebellar infarction. The authors advanced the view that critical PLC lesions are located in the cerebro-ponto-cerebellar pathways. Following disruption of these pathways, the cerebellum, which is crucially involved in adjusting the execution of crying and laughing to its appropriate cognitive and social context, only receives partial information about the context. As a result, the cerebellum operates on the basis of incorrect contextual data which leads to inadequate and even chaotic behaviour. This alternative hypothesis is in accordance with the behavioural-affective disturbances, including emotional fluctuations ranging from disinhibited giggling to inconsolably crying, found in children and adults with CCAS (Levisohn et al., 2000; Schmahmann and Sherman, 1998). Dimova et al. (2009) reported a child who presented with transient mutism and pathological laughter in the context of cerebellitis. This patient frequently reacted in a dramatic way with outburst of intense PLC during the follow-up period of 11.5 months. Disrupted emotional and chaotic reactions were extremely difficult to regulate and were entirely inappropriate with regard to the social or emotional context. Our patient seems to be the first in whom the phenomenon of PLC is described in the context of PFS. In addition, our findings fully support the recently acknowledged role of the cerebellum in the contextual regulation of emotions and affect.

3.5. *Postoperative cognitive decline and persistent behavioural and emotional disturbances*

Postoperative neurocognitive and behavioural-affective symptoms have only been scantily documented in adult patients with the PFS and no long-term neurocognitive follow-up studies have been conducted (see Appendix). Indeed, aside from general memory disturbances in two patients [1, 18] oral apraxia in three cases [6, 18, 20], attentional disturbances in two patients [17, 18] and anomia in one case [18] no information about the postoperative cognitive status of adult patients with PFS is provided. At the behavioural-affective level, lethargy [1], emotional aspontaneity [10], depressed mood [17, 18] and emotional lability [18, 20, 21] were observed in a limited number of patients.

The lack of formal neurocognitive and behavioural follow-up data in adult patients with postoperative PFS might imply that a number of clinically relevant symptoms may have been left unnoticed and untreated. In this patient a generalised cognitive decline, reflected by defective verbal and performal IQ levels, prominent executive dysfunctions, decreased immediate memory levels, attentional deficits, disrupted language dynamics, disturbed confrontational naming and ataxic dysarthria, persisted during longitudinal follow-up. When the patient was referred on an inpatient basis to a neurological rehabilitation centre 11.5 months after tumour surgery a wide range of invalidating behavioural and affective abnormalities were still present.

3.6. *Hydrocephalus as a possible risk factor*

In the early reports meningitis and hydrocephalus were considered the most likely causes of postoperative cerebellar mutism in children with posterior fossa tumours (e.g., Humphreys, 1989; Ferrante et al., 1990). Later studies, however, demonstrated no such association (e.g., Crutchfield et al., 1994; Van Calenbergh et al., 1995; Gelabert-Gonzalez and Fernandez-Villa, 2001). In a series of 41 children with the PFS, Catsman-Berrevoets and Aarsen (2010) did not find a correlation between PFS and pre- and postoperative hydrocephalus. Davis et al. (2011) investigated the impact of hydrocephalus on the development of cognitive, motor, academic and attentional skills in 15 children who underwent cerebellar tumour surgery. Group analyses revealed that patients with moderate hydrocephalus obtained higher scores on various cognitive and attentional tasks than patients with no or severe hydrocephalus. As such no consistent impact of the degree of hydrocephalus was found on neurodevelopmental outcome measures. Comparison of patients with and without a shunt treatment revealed that patients with a shunt performed better on various cognitive tests but not on motor tasks. However, since most patients with a shunt belonged to the 'moderate group', shunt treatment and severity of hydrocephalus may be confounding factors. Overall, neither severity of hydrocephalus nor shunt treatment appeared to be reliable predictors of cognitive and motor outcome in children with cerebellar tumours.

Hydrocephalus in our patient was treated with a permanent ventriculoperitoneal shunt 24 days after the operation. Pre- and postoperative hydrocephalus and increased intracranial pressure might have negatively influenced the clinical course. However, although repeat CT scans demonstrated rapid normalisation of the ventricular volumes after the shunt was installed the patient's clinical condition progressively worsened. Seven days after treatment of the hydrocephalus a state of akinetic mutism was reached and CCAS persisted during follow-up.

Table 4 – Group characteristics of the adult and paediatric patient population.

	Children		Adults	
	Mean or percentage (number of patients)	SD	Mean or percentage (number of patients)	SD
Age	7.4 years	3.2	40.3 years	19.4
Male	59.2% (100/169)		66.7% (14/21)	
Female	40.8% (69/169)		33.3% (7/21)	
Aetiology				
Tumoural	98.5% (196/199)		85.7% (18/21)	
Medulloblastoma	67.8% (135/199)		28.6% (6/21)	
Astrocytoma	20.6% (41/199)		9.5% (2/21)	
Ependymoma	10.1% (20/199)		0	
Haemangioblastoma	0		14.3% (3/21)	
Carcinoma	0		14.3% (3/21)	
Papilloma	0		4.8% (1/21)	
Gangliocytoma	0		9.5% (2/21)	
Vascular	1.5% (3/199)		14.3%	
AVM	1% (2/199)		9.5% (2/21)	
Haematoma	.5% (1/199)		4.8% (1/21)	
Lesion site				
Midline	90.7% (166/183)		57.2% (12/21)	
Midline + RH/LH	9.3% (17/183)		/	
Midline + RH	0		9.5% (2/21)	
Midline + LH	0		4.8% (1/21)	
RH	0		9.5% (2/21)	
LH	0		9.5% (2/21)	
4th ventricle	0		9.5% (2/21)	
Latency mutism	1.5 days (150)	1.7	2.3 days (21)	3.0
Duration of mutism	49.7 days (145)	85.5	38.2 days (35)	35.0
Dysarthria	98.8% (165/167)		100% (18/18)	

AVM = arteriovenous malformation; RH = right cerebellar hemisphere; LH = left cerebellar hemisphere.

3.7. *Involvement of the cerebello-cerebral network as evidenced by SPECT*

CCAS (Schmahmann and Sherman, 1998) reflects functional disruption of the cerebello-cerebral network. In this patient, pivotal involvement of this network is demonstrated by means of a correlation between SPECT and the pre- and postoperative clinical findings. During the phase of akinetic mutism, aggravation and marked extension of the perfusional deficits in the prefrontal brain regions was found. This finding is consistent with Denays et al. (1994) who related bilateral mediofrontal hypo-activity on SPECT to akinetic mutism, motor disability and incontinence. In addition, clinical improvement was associated with restored perfusion bilaterally in the frontal lobe as well as in the right cingulate gyrus. Remission of akinetic mutism was reflected by a normalisation of cerebral blood flow in the inferior frontal region. The phenomenon of cerebello-cerebral diaschisis in our patient adds to the pathophysiological hypothesis that PFS might result from decreased transmission of excitatory impulses from the deep nuclei of the cerebellum through the dentatothalamic connections to the cortical areas crucially involved in cognition and behavioural and affective regulation (Mariën et al., 2001, 2003, 2009; Catsman-Berrevoets and Aarsen, 2010; Miller et al., 2010).

3.8. *PFS in adults and children*

Table 4 shows a comparison of group characteristics for the adults and children. From the very limited information available on the PFS in the adult population it can be concluded that the phenomenology of the syndrome is quite similar to that in children in that mutism and behavioural-affective abnormalities typically develop after a symptom-free interval following surgery and that the duration of the mutistic period is similar.

The discrepancy in prevalence of PFS in adults and children may be related to the fact that posterior fossa tumours are more common in children. Indeed, up to 54–70% of paediatric brain tumours are located in the posterior fossa (O'Brien et al., 2001; Strother et al., 2002), whereas in adults they only account for 15–20% of the tumours (Sklar et al., 2004). In addition, it has been suggested that medulloblastomas – which are the most common malignant central nervous tumour in children accounting for 20–40% of paediatric brain tumours (Menon et al., 2008; Akyüz et al., 2008; Küpeli et al., 2011) – are negatively associated with the occurrence of cerebellar mutism (67.8% after surgery for a cerebellar medulloblastoma, Table 4) and neuropsychological outcome (Catsman-Berrevoets et al., 1999; Ronning et al., 2005). By contrast, medulloblastomas only account for 1% of the brain tumours in adults (Menon et al., 2008).

The postoperative latency of 31 days for the development of mutism in our patient is substantially longer than the mean of 2.3 days (range 0–14 days, SD 3) found in the adult population and also substantially extends the mean of 1.5 days recorded in the paediatric patient group (range 0–11 days, SD 1.7 days) (De Smet et al., 2007). The difference in latency of mutism in children and adults is not significant (Student *T*-test, $p = 1.615$). The duration of akinetic mutism in our patient agrees with the duration of mutism found in the adult patient group (range 1–120 days, mean 38.2 days, SD 35). Although the duration of mutism varies in adults and children (half-day to 2.5 years, 48.6 days, SD 83.3) (De Smet et al., 2007) this difference does not reach significance (Student *T*-test, $p = .623$). With respect to the speech characteristics following the period of mutism in adults, it appears that speech deficits were present in all patients when information was available. Follow-up information, varying between 2 months and 4 years post-surgery was provided for 14 of the 21 patients. Six of these patients (43%) still presented impaired speech at last follow-up.

Similar as in our patient, a broad spectrum of clinically relevant linguistic, cognitive and behavioural-affective sequelae have been identified in the paediatric patient population. These symptoms may have a negative impact on scholastic achievements and cognitive and affective development throughout lifespan (Levisohn et al., 2000; Pollack, 1997; Scott et al., 2001; Siffert et al., 2000; Steinbok et al., 2003; Steinlin et al., 2003; Aarsen et al., 2004; Ronning et al., 2005; Catsman-Berrevoets and Aarsen, 2010; De Smet et al., 2009). Ronning et al. (2005), for instance, found persistent cognitive dysfunctions in young adults treated in childhood for posterior fossa tumours. Impaired scores were found on attention, executive functioning and measures of motor speed (Ronning

et al., 2005). Although significant effects of chemo- and radiotherapy should be taken into consideration when long-term effects are studied (Docking et al., 2007), it is unlikely that radiation treatment alone can be regarded as the sole contributor to the persistence of long-term cognitive deficits. For example, De Smet et al. (2009) reported long-term cognitive and neurobehavioural problems in 2 patients who did not receive radiation treatment. As a result, follow-up studies in children have convincingly shown that the PFS has a far less favourable outcome than initially expected (Van Dongen et al., 1994; Ersahin et al., 1996; Catsman-Berrevoets et al., 1999) and may have significant repercussions on quality of life throughout lifespan.

4. Concluding remarks

In conclusion, pre- and postoperative cognitive, behavioural and SPECT findings in an adult patient are reported who developed PFS with an atypical latency and unique postoperative clinical evolution. In the preoperative phase, clinically relevant cognitive, behavioural and affective abnormalities were identified, matching a diagnosis of CCAS. Cerebellar and brainstem symptoms accompanied by prefrontal-like behavioural and affective abnormalities, peduncular hallucinations and confusion evolving to psychosis characterised the immediate postoperative course. Full-blown akinetic mutism subsequently developed, lasted for 12 days and then alternated with episodes of diminished responsiveness in which the rare phenomenon of PLC occurred. From a semiological point of view "relapsing-remitting akinetic mutism" and PLC in our patient add to current insights in the clinical expression and symptom variability of the PFS in adults. Akinetic mutism resolved after treatment with a non-ergoline dopamine-agonist but CCAS persisted during longitudinal follow-up. As evidenced by a close parallelism between SPECT and clinical findings, CCAS as well as PFS seem to reflect functional disruption of the cerebello-cerebral network crucially involved in the processing of cognitive, behavioural and affective functions. These findings may indicate that both syndromes not only share overt semiological resemblances (basically differing in extent and severity of symptoms), but also a common pathophysiological substrate. From the very limited information available on the PFS in the adult population it can be concluded that the phenomenology of the syndrome is quite similar to that in children in that mutism and behavioural-affective abnormalities typically develop after a symptom-free interval following surgery and that the duration of the mutistic period is similar. Furthermore, it can be concluded that in the few patients who have been described there is a lack of information on both the preoperative and postoperative cognitive, behavioural and emotional symptoms.

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Appendix

Reference and patient number	Age	Sex	Aetiology	Lesion site	Latency of mutism (days)	Duration of mutism (days)	Cognitive symptoms	Behavioural-affective symptoms	Speech symptoms	Follow-up symptoms (time)
Tumoural aetiology										
1. Moore (1969)	68	M	Haemangioblastoma	RH	1	30	Memory	Lethargy	Thick	Monotone (6 m); normal (1.8 y)
2. Salvati et al. (1991)	20	M	Medulloblastoma	Vermis	2	28	?	?	Dysarthria	Dysarthria (6 m)
3. D'Avanzo et al. (1993)	45	M	Medulloblastoma	Vermis + RH	2.5	60	?	?	Cerebellar dysarthria with scanning speech	Scanning speech (3 m)
4. D'Avanzo et al. (1993)	20	F	Medulloblastoma	Vermis	3	42	?	?	Scanning speech	Scanning speech (5 m)
5. Çakir et al. (1994)	61	M	Carcinoma	RH	0	4	?	?	?	?
6. Dailey et al. (1995)	20	F	Astrocytoma	Vermis	0.5	56	Oral pharyngeal apraxia	?	?	?

7.	Bhatoe (1997)	28	M	Haemangioblastoma	LH	3	7	?	?	Near normal speech	
8.	Kai et al. (1997)	71	M	Haemangioblastoma	Vermis	2	28	?	?	Dysarthria	Normal (4 m)
9.	Kai et al. (1997)	74	F	Carcinoma	Vermis	2	21	?	?	Mild dysarthria	Almost normal (2 m)
10.	Caner et al. (1999)	18	F	Choroid plexus papilloma	4th vent	6	?	?	Aspontaneity	?	Normal (6 m)
11.	Ildan et al. (2002)	32	M	Medulloblastoma	Vermis	2	28	?	?	Dysarthria	Fluent (4 m)
12.	Ildan et al. (2002)	44	M	Astrocytoma	Vermis	2	21	?	?	Dysarthria	Mild dysarthria (9 m)
13.	Sajko et al. (2004)	62	M	Medulloblastoma	Vermis	2	?	?	?	Dysarthria	Died (1 m)
14.	Adachi et al. (2005)	18	F	Mixed neuronal-gial	4th vent	14	15	?	?	?	Normal (4 y)
15.	Sherman et al. (2005)	56	F	Carcinoma (+Haematoma)	LH	1	1	?	?	Ataxic dysarthria (slurred)	Normal (1 y)
16.	Akhaddar et al. (2008)	22	M	Medulloblastoma	Vermis	0	10	?	Normal	Dysarthria	Normal (4 y)
17.	Afshar-Oromieh et al. (2010)	36	M	Gangliocytoma	Vermis + LCB	1	120	Attention deficits	Depressive mood, reduced voluntary activity	Dysarthria	?
18.	Afshar-Oromieh et al. (2010)	31	F	Gangliocytoma	Vermis	2	?	Involuntary orofacial movements; memory and attention deficits; anomia	Emotional lability, depression, reduced voluntary activity	Dysarthria	?
Vascular aetiology											
19.	Dunwoody et al. (1997)	53	M	Arteriovenous malformation	Vermis	1	21	?	?	Dysarthria, staccato	Normal (2 m)
20.	Coplin et al. (1997)	47	M	Haematoma	Vermis	0	105	Bucco-lingual dyscoordination	Emotional lability	Ataxic dysarthria	Ataxic dysarthria (9 m)
21.	Idiaquez et al. (2011)	20	M	Arteriovenous malformation	Vermis + RCB	2	90	?	Irritability and labile affect	Dysarthria	?

M = male; F = female; RH = right cerebellar hemisphere; LH = left cerebellar hemisphere; 4th vent = 4th ventricle; m = month(s); y = year(s).

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