Neuropsychological consequences of cerebellar tumour resection in children
Cerebellar cognitive affective syndrome in a paediatric population

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Summary
Acquired cerebellar lesions in adults have been shown to produce impairments in higher function as exemplified by the cerebellar cognitive affective syndrome. It is not yet known whether similar findings occur in children with acquired cerebellar lesions, and whether developmental factors influence their presentation. In studies to date, survivors of childhood cerebellar tumours who demonstrate long-term deficits in cognitive functions have undergone surgery as well as cranial irradiation or methotrexate treatment. Investigation of the effects of the cerebellar lesion independent of the known deleterious effects of these agents is important for understanding the role of the cerebellum in cognitive and affective development and for informing treatment and rehabilitation strategies. If the cerebellar contribution to cognition and affect is significant, then damage in childhood may influence a wide range of psychological processes, both as an immediate consequence and as these processes fail to develop normally later on. In this study we evaluated neuropsychological data in 19 children who underwent resection of cerebellar tumours but who received neither cranial irradiation nor methotrexate chemotherapy. Impairments were noted in executive function, including planning and sequencing, and in visual–spatial function, expressive language, verbal memory and modulation of affect. These deficits were common and in some cases could be dissociated from motor deficits. Lesions of the vermis in particular were associated with dysregulation of affect. Behavioural deficits were more apparent in older than younger children. These results reveal that clinically relevant neuropsychological changes may occur following cerebellar tumour resection in children. Age at the time of surgery and the site of the cerebellar lesion influenced the neurobehavioural outcome. The results of the present study indicate that the cerebellar cognitive affective syndrome is evident in children as well as in adults, and they provide further clinical evidence that the cerebellum is an essential node in the distributed neural circuitry subserving higher-order behaviours.

Keywords: cerebellum; cognition; affect; behaviour; cerebellar cognitive affective syndrome; posterior fossa syndrome

Abbreviations: EOWPVT-R = Expressive One-Word Picture Vocabulary Test–Revised; VMI = Visual Motor Integration; WISC-R = Wechsler Intelligence Scale for Children—Revised; WPPSI-R = Wechsler Preschool and Primary Scales of Intelligence—Revised; WSC III = Wechsler Intelligence Scale for Children, Third Edition

Introduction
A cerebellar cognitive affective syndrome has been described recently in adults with acquired cerebellar lesions that is characterized by deficits in executive function, spatial cognition, linguistic processing and affect regulation, resulting in overall intellectual impairment (Schmahmann and Sherman, 1998). These findings extend earlier observations of executive (Akshoomoff and Courchesne, 1992; Courchesne et al., 1994), visual–spatial (Botez et al., 1985; Bracke-Tolkmitt et al., 1989; Wallesch and Horn, 1990; Attig et al., 1991), linguistic (Bracke-Tolkmitt et al., 1989; Fiez et al., 1992; Appolonio et al., 1993; Molinari et al., 1997; Silveri et al., 1998) and behavioural (Heath, 1977) changes in adult patients with focal cerebellar damage, and they were subsequently supported by the observations
of Malm and colleagues that central aspects of attention and visual–spatial skills are impaired in young adults with cerebellar infarcts (Malm et al., 1998).

Tumours of the cerebellum and brainstem account for half of all brain tumours in children (Heideman et al., 1993), and the prognosis for survival is improving as a result of advances in treatment. The realization that cerebellar lesions produce clinically relevant intellectual disability makes it important to determine whether neuropsychological abnormalities occur in the long-term survivors of paediatric cerebellar tumours. Little is known about the neuro-behavioural sequelae resulting specifically from the resection of these tumours in this population. The impairments in intelligence, memory, language, attention, academic skills and psychosocial function that have been reported (Waber, 1990; Dennis et al., 1996) have been observed in groups that have undergone not only cerebellar resection but also cranial irradiation and/or chemotherapy. This makes it difficult to understand the clinical consequences of the cerebellar resection itself because radiation necrosis is associated with deficits in general intelligence, academic achievement, verbal knowledge and reasoning, and perceptual motor abilities, and methotrexate also causes substantial neurological and neurobehavioural impairment (Duffner et al., 1983; Packer et al., 1989; Dowell et al., 1991; Glauser and Packer, 1991; Radcliffe et al., 1992; Dennis et al., 1996). Among the patients studied by Baron and colleagues, four with astrocytoma did not receive radiation. Two of these showed significant improvement on cognitive and academic measures including mental flexibility and mathematical skill, three were reported to be ‘slow workers’ on teacher rating scales, and all showed significant social or emotional difficulties on the child behaviour checklist (Baron et al., 1988). Packer and colleagues reported a trend for children with astrocytoma who did not receive irradiation to have a better neuropsychological outcome than those with medulloblastoma who did receive irradiation (Packer et al., 1989).

To date there has been no systematic investigation of the long-term neuropsychological consequences in children whose cerebellar tumours have been surgically excised but who have not received cranial radiation or methotrexate. We wished to determine (i) whether such a population would demonstrate neurobehavioural disturbances; (ii) whether the deficits, if present, would resemble the cerebellar cognitive affective syndrome in adults; (iii) whether the deficits would vary according to the site of the lesion; and (iv) whether the deficits would be influenced by developmental issues. This report presents our observations in 19 such children who were studied in order to address these questions.

**Patients and methods**

**Patients**

The records of 48 children who were treated for tumours of the cerebellum at Children’s Hospital, Boston during the 15-year period between 1982 and 1997 and who were referred for subsequent neuropsychological evaluations in the Children’s Hospital Neuropsychology Program were reviewed independently by two investigators (L.L. and Aida Kahn) who were blinded to the results of neuroimaging scans. Patients were included in this study if they had neuropsychological testing prior to radiation or had astrocytomas that did not require radiation, and they were tested during the first 2 years after surgery. Children were excluded if they were too young to receive standard neuropsychological tests, if they were thought not to have recovered from surgery at the time of their baseline neuropsychological evaluation, if they had unusually complicated medical histories, or if they had pre-existing cognitive impairment. Premorbid testing was not available in this group of subjects, but parent reports and school records revealed no areas of concern prior to the onset of the neurological or neurobehavioural symptoms that prompted the initial clinical evaluations. No children received methotrexate, but those who received cisplatin, CCNU [N-(2-chloroethyl-N-cyclohexyl-N-nitrosourea) glyco-phosphaphamide and vincristine were not excluded as these agents have been shown not to have cognitive or behavioural effects (Packer et al., 1989; Radcliffe et al., 1992; Kao et al., 1994; Seaver et al., 1994; Dennis et al., 1996).

**Neuropsychological tests**

Children aged 3–5 years were administered subtests of the Wechsler Preschool and Primary Scales of Intelligence—Revised (WPPSI-R); Naming vocabulary subtest of the Differential Abilities Scale (DAS); Naming Vocabulary; Taylor children’s stories (Logical Memory Passages for Children); Beery Visual Motor Integration (VMI); and the Gardner Expressive One-Word Picture Vocabulary Test—Revised (EOWPVT-R). The WPPSI-R does not include Digit Span, so the younger patients did not receive this test. Children aged 5–16 years were administered subtests of the Wechsler Intelligence Scale for Children—Revised (WISC-R) or Third Edition (WISC III); Automatized Series; Boston Naming Test; Sentence Memory and Story Memory of the Wide Range Assessment of Memory and Learning; Beery VMI; and the Rey–Osterrieth Complex Figure. Children aged 12–16 years were given the Verbal Fluency, Repeated Patterns and Stroop colour and word Tests in addition to all tests given to the 5- to 16-year-old children. The Grooved Pegboard test was used with all ages as a measure of fine motor speed and coordination. Given the retrospective nature of this study, not all tests were administered to all patients.

**Definitions of deficit**

Expressive language or visuospatial functions were considered impaired if scores were ≥1.5 SD below the mean. Verbal memory was considered impaired if a test of story memory was ≥1.5 SD below the mean. A deficit in Digit span...
Span alone was not considered a sufficient criterion for a deficit in verbal memory because children who scored poorly on this subtest tended to have difficulties with sequencing and planning of verbal output rather than memory per se. The Rey–Osterrieth Complex Figure recall condition was not considered a sufficient criterion for measuring visual memory because poor initial organization of the figure on the copy condition would confound memory performance for many of the children.

Measures of behavioural/affective regulation
Behavioural or affective disturbance was assessed with the Behavioural Rating Scale, Achenbach’s Child Behaviour Checklist and parents’ reports. Patients were classified as having a deficit in affect regulation if this was a persistent postoperative change documented by a clinician and verified by parents. Patients with increased irritability preceding tumour diagnosis and resection were not considered to have affective disturbance if the irritability resolved postoperatively. Patients were classified as having posterior fossa syndrome if they exhibited a resolving, postoperative mutism (from 2 to 48 h) with or without irritability and impairment of volitional activities. Behavioural disturbances were not considered to be persistent if they occurred as part of the posterior fossa syndrome and resolved as the mutism cleared.

MRI analysis
The postoperative MRI films for each case were analysed according to the atlas of Schmahmann and colleagues, which permits the reliable identification of cerebellar fissures and lobules (Schmahmann et al., 1999). In two patients, lesion localization was based on CT scans because MRI scans were not available. For all cases, the lesion was assigned to one or more of the following categories: vermis (minimal, or split vermis versus extensive vermis damage); deep cerebellar nuclei (left versus right); hemisphere (posterior lobe versus anterior lobe; right versus left hemisphere). The lack of fine detail in some of the clinical scans and the distortion of tissue by the surgical ablations in others limited our ability to analyse all the lesions according to the cerebellar lobules affected. Where there was sufficient detail, the lobules affected were described beyond the more general characterization of the region of cerebellum involved. All the images were analysed (by J.D.S.) blind to the patient’s identity and neuropsychological data.

Results
Patient data
Of the 48 patients screened, 34 had medulloblastoma, seven astrocytoma and seven ependymoma.

The study group consisted of 19 children. Eleven had medulloblastoma, seven astrocytoma and one an ependymoma. The children ranged in age from 3 years 3 months to 14 years 10 months (mean age 8 years 2 months) at the time of tumour resection, which occurred 1–2 weeks after diagnosis. The time between surgery and neuropsychological testing ranged from 1 to 22 months (mean 5.1, SD 6.4 months). Ages of each child at diagnosis and neuropsychological evaluation are presented in Table 1. Eight children received chemotherapy prior to neuropsychological testing. Two children (cases 13 and 19) had evidence of dural metastasis without invasion of the brain parenchyma. Two cases (cases 13 and 15) had hydrocephalus that required shunting.

Neuropsychological findings
There was agreement between the two reviewers in all cases. Summary data regarding the presence or absence of neuropsychological deficit and lesion localization are presented in Table 1. Table 2 shows individual scores for each participant in a subset of tests consisting of those that were most consistently administered to all 19 children. No consistent differences were observed between children tested within the first 3 months after surgery and those tested up to 22 months after surgery.

Three patients met qualitative rather than quantitative criteria for deficit in a particular domain, because they were administered only one test in that domain in which they performed at least 1.5 SD below the norm or exhibited behavioural deficits that were not captured by absolute test scores on at least two tests. Standardized Intelligence Scale subtests did not always reflect the extent of the clinical and neuropsychological deficits in the children. Case 5 illustrates this point, in that the Block Design and Picture Arrangement subtests of the WISC-R that assess visual–spatial function were normal, but the Rey–Osterrieth copy and delayed recall were highly abnormal (Figs 1 and 2).

Cognition
Seven of the 19 patients (37%) met criteria for expressive language deficits. Another four (cases 4, 7, 18 and 19) demonstrated word-finding difficulties detected either in testing (difficulty naming objects in the Picture Completion test) or in spontaneous conversation, in the context of otherwise intact language abilities. Seven (37%) had deficits in visual–spatial functions. Three (16%) had deficits in both expressive language and visual–spatial functions. In addition to these deficits in expressive language and visual–spatial functions, eight out of the 14 (57%) children who were administered the Digit Span test performed poorly. Of these eight, five had below-average age-scaled scores, ranging from 5 to 8 (where 10 is average) and two had poor scores relative to their own scaled scores on other subtests (cases 5 and 6). Some with average scores on Digit Span (e.g, case 11) showed perseveration and difficulty establishing set. Five of 15 tested (33%) had verbal memory deficits along with other deficits in visual–spatial and/or language function.
Table 1 Patient data

<table>
<thead>
<tr>
<th>Case</th>
<th>Age at resection (years)</th>
<th>Age at evaluation (years)</th>
<th>Language</th>
<th>Visual–spatial function</th>
<th>Visual–spatial memory</th>
<th>Affect</th>
<th>Executive function</th>
<th>Pathology</th>
<th>Tumour site</th>
<th>Chemo-therapy</th>
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<tr>
<td>1</td>
<td>4–0</td>
<td>4–02</td>
<td>–</td>
<td>–</td>
<td>n.t.</td>
<td>–</td>
<td>–</td>
<td>Medulloblastoma</td>
<td>Inferior vermis, (minimal)</td>
<td>–</td>
</tr>
<tr>
<td>2</td>
<td>14–10</td>
<td>16–06</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>Astrocytoma</td>
<td>Mostly right posterior lobe, paravermal to lateral</td>
<td>–</td>
</tr>
<tr>
<td>3</td>
<td>3–06</td>
<td>4–04</td>
<td>–</td>
<td>–</td>
<td>n.t.</td>
<td>–</td>
<td>–</td>
<td>Medulloblastoma</td>
<td>Left anterior lobe, rostral posterior; right paravermal</td>
<td>–</td>
</tr>
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<td>4</td>
<td>9–09</td>
<td>9–10</td>
<td>†</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>Medulloblastoma</td>
<td>Left vermis and deep nuclei; atrophy right crus I, II</td>
<td>+</td>
</tr>
<tr>
<td>5</td>
<td>6–0</td>
<td>6–11</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>Astrocytoma</td>
<td>Vermis, left hemisphere (anterior, posterior lobes)</td>
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</tr>
<tr>
<td>6</td>
<td>5–11</td>
<td>6–03</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Astrocytoma</td>
<td>Left nuclei, posterior lobe white matter, paravercial</td>
<td>–</td>
</tr>
<tr>
<td>7</td>
<td>5–03</td>
<td>5–04</td>
<td>†</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Astrocytoma</td>
<td>Left posterior lobe</td>
<td>–</td>
</tr>
<tr>
<td>8</td>
<td>7–04</td>
<td>7–06</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>+</td>
<td>Medulloblastoma</td>
<td>Midline nuclei, not dentate; enlarged 4th ventricle</td>
<td>+</td>
</tr>
<tr>
<td>9</td>
<td>4–10</td>
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<td>†</td>
<td>–</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>Medulloblastoma</td>
<td>Entire verm</td>
<td>+</td>
</tr>
<tr>
<td>10</td>
<td>3–03</td>
<td>3–08</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Astrocytoma</td>
<td>Vermis, left and right medial nuclei but not dentate</td>
<td>–</td>
</tr>
<tr>
<td>11</td>
<td>13–0</td>
<td>13–02</td>
<td>†</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>Medulloblastoma</td>
<td>Right verm, paravermal, medial nuclei, not dentate</td>
<td>+</td>
</tr>
<tr>
<td>12</td>
<td>3–07</td>
<td>3–09</td>
<td>–</td>
<td>–</td>
<td>n.t.</td>
<td>–</td>
<td>–</td>
<td>Ependymoma</td>
<td>Paravermal (split verm); enlarged 4th ventricle</td>
<td>–</td>
</tr>
<tr>
<td>13</td>
<td>11–09</td>
<td>12–0</td>
<td>†</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>Medulloblastoma</td>
<td>Right verm, right and left nuclei</td>
<td>+</td>
</tr>
<tr>
<td>14</td>
<td>6–11</td>
<td>7–0</td>
<td>+</td>
<td>–</td>
<td>n.t.</td>
<td>–</td>
<td>+</td>
<td>Medulloblastoma</td>
<td>Right nuclei and medial dentate; verm</td>
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</tr>
<tr>
<td>15</td>
<td>10–0</td>
<td>10–02</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>+</td>
<td>Medulloblastoma</td>
<td>Left posterior lobe; verm not nuclei; enlarged 4th ventricle</td>
<td>+</td>
</tr>
<tr>
<td>16</td>
<td>13–09</td>
<td>15–06</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Astrocytoma</td>
<td>Left inferior verm, not nuclei; left lobules VIIB and VIII</td>
<td>–</td>
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<tr>
<td>17</td>
<td>11–10</td>
<td>12–01</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>Medulloblastoma</td>
<td>Entire verm</td>
<td>+</td>
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<tr>
<td>18</td>
<td>5–03</td>
<td>5–05</td>
<td>†</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>Medulloblastoma</td>
<td>Right verm, paravermal; medial crus II, VII, VII</td>
<td>+</td>
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<tr>
<td>19</td>
<td>11–05</td>
<td>11–06</td>
<td>†</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Medulloblastoma</td>
<td>Split verm, minimal destruction</td>
<td>–</td>
</tr>
</tbody>
</table>

+ = Abnormal; – = normal; † = language impairment restricted to word finding difficulties; n.t. = not tested.

Affect
Six patients (32%) had deficits in affect regulation, and they all had extensive vermis damage. In contrast, none of the patients without affect problems had extensive vermis damage. Five of nine children (56%) who sustained extensive vermis damage exhibited the posterior fossa syndrome postoperatively. Thus, children with extensive vermis damage were more likely to exhibit changes in affect regulation than children with an intact vermis or only minimal vermis damage.

Motor functioning
Fourteen of 19 patients (74%) had mild to marked impairment in fine motor coordination. Four of the five patients who did not have fine motor deficits were also free of cognitive deficits. Four patients with fine motor deficits did not have cognitive deficits.

Age effects
Patients who were youngest at the time of testing were least likely to show neuropsychological deficits. Of the nine patients who were younger than 7 years, only three (33%) showed any deficit, whereas eight of the 10 patients (80%) older than 7 years had deficit. These age effects were confounded by tumour type. The majority of younger children (75%) had tumours other than medulloblastoma (i.e. astrocytoma or ependymoma).

Functional topography
Damage was localized to the left cerebellar hemisphere in four patients and the right hemisphere in two. Both patients with lateralized right cerebellar damage but without extensive vermis damage had language deficits with intact visual-spatial function. One of the four cases with left cerebellar damage had visual-spatial deficits with intact language function. The remaining three cases with left cerebellar damage had no deficit in either language or visual-spatial function. The cases with damage to the cerebellar hemispheres were too few and varied for us to be able to identify with certainty the patterns of injury within the posterior and anterior lobes.
Table 2 Neuropsychological scores in a subset of tests most consistently administered to all 19 children (average age is 10 years, SD = 3)

<table>
<thead>
<tr>
<th>Case</th>
<th>Block design</th>
<th>Picture arrangement</th>
<th>Vocabulary</th>
<th>Information</th>
<th>Digit span</th>
<th>Fine motor deficit</th>
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Representative case studies
Cases 5, 9 and 15 are presented in more detail below to illustrate the range of deficit profiles. The cases coincidentally varied with respect to age, sex and treatment.

Case 5
Six-year-old right-handed male. Left cerebellar cystic astrocytoma. Resected at 6 years 0 months (Fig. 1). Age at baseline evaluation, 6 years 11 months. Overall intellectual functioning average (VIQ = 105, PIQ = 100, FSIQ = 102, where VIQ, PIQ and FSIQ are verbal, performance and full-scale IQ, respectively). Quality of responses in verbal tasks indicated the small sample size in this retrospective record review that premorbid skills were high average (excellent vocabulary) made it difficult to control for variables that could affect cognitive outcome such as chemotherapy, hydrocephalus and ventriculoperitoneal shunting. Two patients had dural metastases, although neither had focal deficits attributable to these lesions. The two patients with hydrocephalus required a ventriculoperitoneal shunt via a right parietal approach; one had deficits in visual–spatial skills and affect, and the other had deficits in language and verbal memory, making it unlikely that these findings were attributable to the shunt surgery. Eight children received vincristine, cyclophosphamide or cisplatin. These have been shown not to have adverse cognitive effects (Dennis et al., 1996), but the potential contribution of these treatment variables to the cognitive changes is acknowledged. Standard measures of executive(prefrontal function in this group were limited, and included the Stroop test and assessments of verbal fluency. The lack of in-depth analyses of executive function in the records limited the strength of the conclusions.

Language. Speech slow but well articulated. Language processing slow. Scores on verbal tests were average to above average. Subtle problems with retrieval on the Boston Naming Test (facilitated with phonemic cues) and word-finding difficulty in conversation. Unable to inhibit inappropriate responses and naming difficulties on Picture Completion. Perseveration on WISC-R subtests, with intrusion of elements from previous items, and difficulty giving alternate answers.

Visual–spatial. Problems processing and integrating information on visual–spatial tasks, such as Block Design,
on the left following surgery and included dysmetria and dysdiadochokinesia.

**Follow-up data.** Behavioural and attentional problems and slow academic progress persisted. Eight years after surgery, writing was slow and troublesome. Spontaneous language was notable for hesitancy, markedly impaired fluency and difficulty in word-finding. Subtle syntax errors were also noted. He had long response latencies, tended to miss important details, and often did not recognize his errors.

**Case 9**

Five-year-old right-handed male. Midline medulloblastoma resected at 4 years 10 months (Fig. 3). Ventriculoperitoneal shunt followed by vincristine, cyclophosphamide and cisplatin. Age at baseline evaluation 5 years 0 months. General intellectual function average for Performance subtests (PIQ = 98) and Mildly Mentally Deficient for Verbal subtests (VIQ = 67), with high variability.

**Language.** Spontaneous language impaired in quality, organization and level of information. Articulation was dyspraxic, dysarthric and agrammatic. Spontaneous vocabulary and word retrieval were below age level. He often pointed instead of naming objects, and spoke in sentence fragments or phrases. Difficulty with formulation of responses, giving perseverative or associative responses. Comprehension was impaired. Immediate repetition of sentences contained paraphasic, grammatical and phonemic processing errors, although simple items were repeated correctly, suggesting adequate auditory perception. Confrontation naming (EOWPVT-R) was significantly below age expectations, with incorrect responses usually in the correct semantic category. Difficulty naming details on Picture Completion. Receptive vocabulary (Peabody Picture Vocabulary Test) and WPPSI-R verbal subtest scores well below average.

**Verbal memory.** Immediate recall of sentences displayed language processing errors with functional immediate memory for limited amounts of verbal information. In contrast, he could not recall the paragraph-long Taylor Children’s Stories, indicating difficulty sustaining attention when overloaded with information.

**Visual–spatial.** Difficulty orienting oblique figures and discriminating figure from ground on Block Design. Analysis of visual details within context was quick and accurate on Picture Arrangement, but there was difficulty naming details. Initial inattention to right hemispace on Animal Pegs. Performance on VMI significantly below age expectations. Drawings developmentally delayed, with graphomotor imprecision. Without a motor component, visual perceptual skills were average.

**Behaviour/affect.** Mood was labile and irritable, shifting
frequently between anger and crying. Marked difficulty modulating behaviour and sustaining task performance on verbal subtests.

**Motor.** Performance on timed motor tests was below age level. Graphomotor control was poor.

**Case 15**
Ten-year-old right-handed female. Midline medulloblastoma, resected at age 10 years 0 months. Ventriloperitoneal shunt, chemotherapy with vincristine and cisplatin. Age at baseline evaluation 10 years 2 months. General intellectual function average (estimated FSIQ = 105) with difficulties in visual–spatial, constructional skills and motor control.

**Language.** Mute postoperatively, but recovered verbal functioning 7 weeks before baseline neuropsychological evaluation. Spontaneous conversation, comprehension, and confrontation naming with some semantic paraphasias. Impaired repetition of complex grammatical constructions. Scores on WISC-R verbal subtests high average.

**Verbal memory.** Worse on tests requiring exact repetition. Difficulty repeating sentences verbatim (Sentence Memory), but above average recall of longer stories (Logical Memory) that required less emphasis on exact repetition. Repeated only four digits forwards, three digits backwards.

**Visual–spatial.** Difficulty with all tasks of visual perception and construction, showing incomplete appreciation of figural relationships and configurations. Drawings of the Rey–Osterrieth figure were anomalous: basic shape preserved but figure drawn in an unusual, segmented fashion with loss of internal configuration on both copy and memory conditions. Block Design below average, showing difficulty in visual–spatial analysis. In a written narrative based on pictures she omitted words without noticing until asked to read the narrative aloud.

**Behaviour/affect.** She was silly, with disinhibited giggling, but was likable and friendly.

**Motor.** Motor output was slow bilaterally. Trouble sustaining and fine-tuning all graphomotor tasks.

**Discussion**
This is the first systematic study of cerebellar resections in children that focuses on the neuropsychological and behavioural consequences, and that is not contaminated by the use of radiation therapy or methotrexate. The results indicate that children with cerebellar tumour resections show impairments in some or all of a characteristic set of behaviours, including problems with visual–spatial functions, language, sequencing, memory and the regulation of affect.

**Cognitive impairments**
Visual–spatial difficulties were characterized by impairments in the planning and organizational aspects of the tasks. Deficits in expressive language were characterized by brief responses, lack of elaboration, reluctance to engage in conversation, long response latencies and word-finding difficulties. Several children also exhibited difficulties with language initiation even in the context of average scores on verbal tests. Impaired performance on the Digit Span test was not associated with verbal memory problems, and therefore probably reflected deficits in verbal sequencing and planning, or a mental, visual scanning component of this sequencing task. One-third of the children performed poorly on memory tests for stories, some being able to recall only a few details from stories two paragraphs long. Many patients demonstrated difficulty in the initiation of responses and with problem-solving strategies. Deficits in confrontation-naming were ameliorated with...
phonemic clues, and story retrieval improved with multiple-choice prompts. Many patients also failed to organize verbal or visual–spatial material for encoding. These findings extend some of the earlier observations derived from clinical and functional neuroimaging experiments. Poor scores on expressive language tests are consistent with clinical (Bracke-Tolkmitt et al., 1989; Akshoomoff et al., 1992; Appolonio et al., 1993; Schmahmann and Sherman, 1998; Silveri et al., 1998) and functional neuroimaging (Petersen et al., 1988, 1989; Petersen and Fiez, 1993) investigations demonstrating cerebellar involvement in expressive language tasks. Difficulty in the sequential ordering of thoughts and actions has been reported in both clinical and neuropsychological studies of adults with cerebellar lesions (Grafman et al., 1992; Botez-Marquard and Botez, 1997; Schmahmann and Sherman, 1998), and the verbal memory impairment is consistent with imaging studies indicating cerebellar involvement in verbal memory tasks (Grasby et al., 1993; Andreason et al., 1995; Cabeza et al., 1997). The phenomenon of poverty of initiation has been observed in children (Pollack et al., 1995) as well as in some adults with the cerebellar cognitive affective syndrome, in whom decreased verbal fluency, sometimes to the point of mutism, was also observed.

Regulation of affect

Our findings support an association between extensive vermis damage and impaired regulation of affect, including irritability, impulsivity, disinhibition, and lability of affect with poor attentional and behavioural modulation. This pattern is consistent with other clinical evidence of a relation between vermis abnormalities and affective disturbance, such as those seen in children with vermial agenesis (Steinlin, 1997), in adults with the cerebellar cognitive affective syndrome, and in the posterior fossa syndrome that develops in ~15% of children who undergo midline cerebellar surgery and that is characterized by transient postoperative mutism as well as intractable whining, emotional lability, withdrawal and apathy (Pollack et al., 1995).

Patients with affective changes also demonstrated cognitive impairment, which was characterized by language deficits in four patients and visual–spatial dysfunction in two. The reverse did not appear to hold, however, because some patients showed cognitive changes but their affect was normal. These findings have relevance for the hypothesis that affect regulation is principally a function of the vermis and fastigial nucleus (Schmahmann, 1991, 1996), but perhaps both the vermis and the cerebellar hemispheres are involved in executive, linguistic and visual–spatial functions.

Effects of laterality

Only three of the six laterialized astrocytoma cases with cognitive deficits showed deficit patterns compatible with crossed cerebrocerebellar connections. Thus, despite the existence of crossed cerebral diaschisis shown by single photon emission computerized tomography in cortical areas contralateral to cerebellar lesions (Attig et al., 1991), the lateralization of cognitive functions within the cerebellum may not be tightly linked with lateralization of the cerebral cortex.

Age effects

The youngest children were least likely to show deficits in cognition or affect. Medulloblastomas were uncommon in the younger children, however, and it is therefore unclear whether young age or tumour type was the more closely associated with the better functional outcome. A pattern of fewer deficits in the younger children could reflect a lack of sensitivity of the testing. Further, the functional domains in which the older children had difficulty, such as the initiation and organization of speech and visual–spatial organizational skills, do not make large developmental gains between the ages of 7 and 11 years (McKay et al., 1994). The lower incidence of impairment in the younger children may also reflect neural plasticity, analogous to the recovery of language following left-hemispherectomy (Woods and Teuber, 1973; Rasmussen and Milner, 1977). This pattern of results, in which a younger age at tumour diagnosis is associated with a better cognitive outcome, is opposite to the pattern of long-term cognitive outcome seen in children treated with cranial irradiation. This may reflect the differential effects of the single surgical approach versus the delayed neuropathological consequences known to be associated with irradiation (Radcliffe et al., 1992; Dennis et al., 1996).

Functional topography

This study was not large enough to provide convincing evidence in favour of the hypothesis of functional topography of cognition within the human cerebellum (Schmahmann, 1991, 1996), but the dissociation of motor and cognitive disturbances in some of the patients is noteworthy. The presence of cognitive impairments in these children provides further support for the functional relevance of the anatomically defined cerebrocerebellar circuits that link associative and paralimbic regions of the cerebral cortex with the cerebellum and that define the cerebellum as an essential node in the distributed neural circuits that subserve higher-order behaviour (Leiner et al., 1986; Middleton and Strick, 1994; Schmahmann and Pandya, 1989, 1997). These pathways have been proposed as the anatomical substrates subserving the cerebellar contribution to cognition and emotion, and, together with the cerebellar corticonuclear microcircuitry (Ito, 1993), have been essential in developing the hypothesis of dysmetria of thought (Schmahmann, 1991, 1996), a proposed fundamental mechanism of the abnormal behaviours resulting from lesions of the cerebellum.
Further psychological relevance

Neuropsychological test scores in the children do not always provide a complete picture of the behavioural changes. In some of our patients, clinical observations indicated deficits that were not adequately captured by test scores. Further, test scores do not necessarily provide a full picture of the implications of the poor performance. The cognitive deficits and dysregulation of affect in our patients could be related in part to the experience of enduring a life-threatening illness and aggressive treatments including surgery. Further, psychological recovery is dependent, in part, on the ability to comprehend the difficulties one is experiencing and to communicate this to others (Herman, 1992). Impaired verbal expression limits a child’s ability to formulate and communicate feelings and can interfere with the ability to cope effectively. Our patients exhibited uncharacteristic giddiness that is not easily attributed to language-related frustration, and two patients with a change in affect had no language impairment. The explanation for the observations in these children is therefore unlikely to be purely psychological, but it is important to recognize the impact that impairments in expressive language and other behavioural impairments can have on their ultimate recovery.

Future studies

The limitations inherent in this retrospective chart review are readily acknowledged, but the study nevertheless provides an initial understanding of the consequences of cerebellar resection for cognition and affect in children. There are important questions remaining that may be addressed in future prospective studies. The correlation of cognitive deficit with the site of the cerebellar lesion in a larger group of patients studied prospectively and in more detail could test the hypothesis concerning the topography of both sensorimotor and cognitive/affective function within the human cerebellum. The nature and severity of executive functional impairments need to be quantified in more detail, as they appear to be frequent in children, and are a central feature of the cerebellar cognitive affective syndrome in adults. It would be valuable to determine whether the observed disturbances of affect in these patients are matched by a deficit in their perception of emotional tone in others. Longitudinal follow-up is needed to establish whether the deficits we have reported are enduring, as was noted in one patient in this series for whom data was available 8 years after surgery.

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