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SPANS-X Case Study

Mr CT: Suspected early-onset Alzheimer's disease

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1.1. Background

Mr CT, a recently retired 61-year-old primary school teacher, was referred for an outpatient neuropsychological assessment by a consultant neurologist/memory clinic at the arrangement of Mr CT's (now adult) children. His children reported that Mr CT had acquired short-term memory difficulties and behavioural changes, noticeable over the past three to six months.

Mr CT was educated to master's degree level in science and had a secondary school teaching qualification. Since his early 30s he had been teaching, and was well regarded in his work, having won awards for contributions to teaching science in secondary education, and having been a mentor to his students. Divorced for 16 years he had previously lived alone in a two-bedroom house in a suburban area near the school he taught. Then, several months after retirement he had to move, which partially prompted the recognition of Mr CT's suspected decreased ability to cope with change. Notably, Mr CT had a first degree relative (his mother) who had early-onset Alzheimer's disease, a known key risk factor of offspring also developing the disease (Skoog & Blennow, 2001).

Referral information

The neurologist's referral stated that Mr CT increasingly forgot to do jobs and errands, experienced word-finding difficulties and became 'flustered' in the presence of others. She requested a baseline cognitive assessment to identify strengths and weaknesses, and particularly to provide an opinion as to whether there appeared to be indications of early-onset dementia or another neurological condition, or whether the recognised changes related to mood or psychological disturbance, and whether further assistance (e.g. Social Services) and/or assessment (e.g. occupational therapy) were required. The referral indicated there were no structural changes evident to Mr CT's brain on a CT brain scan.

Patient's and others' views of difficulties

The Mr CT was seen in the first instance for a single outpatient interview and testing session, which he attended alone, and therefore further collateral history/information was only available from the neurologist's referral. Mr CT admitted that he found remembering things and keeping himself organised more difficult over the last several months, but said that he did better when alone, when he could make lists, take his time and did not have 'performance' pressure with others around, or his adult children "hassling" him.

He attributed the difficulties of the last few months to the fact he had had to move to a new house, which led to his life and belongings being in disarray, not to mention having to adjust to the changes brought on by retirement. He reported missing the routine and social aspects of going to work every day. Mr CT noted that his children were worried about him because they felt he had changed (cognitively and behaviourally), but he attributed any changes they might have observed to the pressures they put him under when they came to visit him. Historically, Mr CT reported that when faced with difficult challenges in his life, he had always faced them and never really got depressed or anxious. However, recently he said he had been feeling anxious, even panicked, at times, especially during 'stressful events' and when he was 'being watched'. His scores on the Hospital Anxiety and Depression Scale (HADS; Zigmond & Snaith, 1983) indicated slightly elevated (mild) anxiety, but only minimal, non-clinical depression symptoms.

1.2. Integrated observations and SPANS-X score results

Mr CT was pleasant and cooperative, and appeared to put forth exceptional effort and vigilance on all the tasks. When he found tasks difficult, and indeed when he was unable to recall information or answer questions correctly, he tended to voluntarily offer reasons to justify this, such as 'everyone my age finds this sort of thing difficult' and 'I didn't know I was going to have to remember this, so I didn't pay attention' (when in fact he was informed that there would be a delayed memory recall trial, and it was observed that he had concentrated diligently on the learning trial). He demonstrated a few incidents of lapsed attention, appearing distracted on occasion. Halfway through the assessment, Mr CT excused himself for a break, and was shown where to go. He was unable to find his way back to the office, so he caught the attention of another staff member who guided him back to the testing office. He was clearly distressed by this incident and complained that the floor plan was 'very confusing'.

Index level and other assessment interpretation

Supplementary tests

In addition to SPANS-X, it was useful to administer a few supplementary tests, including the National Adult Reading Test (NART; Blair & Spreen, 1989), selected subtests from the Wechsler Adult Intelligence Scale, Third Edition (WAIS-III; Wechsler, 1997a), the Trail Making Test (TMT; Reitan & Wolfson, 1985), Clock Drawing Test (CDT; Rouleau, Salmon, Butters, Kennedy, & McGuire, 1992) and tests of speeded verbal fluency. The NART provided an estimated 'pre-morbid' IQ of 113 (high average, approximately the 81st percentile). His current 'Verbal IQ', as estimated by the WAIS-III, was largely consistent with the NART estimate at 110 (high average, 75th percentile), but his 'Performance IQ' was approximately a standard deviation below this, at 98 (average, 45th percentile). It took 33 seconds for Mr CT to complete Part A of the TMT (50th percentile, average), but he made two 'set loss' errors on Part B, taking 92 seconds overall, equivalent to the low average range in his age group (between the 16th and 25th percentiles) (Strauss, Sherman, & Spreen, 2006). His speeded phonetic verbal fluency (i.e. words that begin with the letters F, A and S) approached expected levels in the high end of the average range (between the 50th and 75th percentiles), but his categorical/semantic fluency (i.e. animals) was again in the low average range (between the 16th and 25th percentiles). He obtained a score of 7/10 on the CDT, with minor distortions in each scoring category, classified as 'mild impairment'. These results began to hint at a level of cognitive decline in some functions, with presumed pre-morbid abilities in the high average range, with several scores dipped into the low average range. From this basis, SPANS-X A was then administered.

SPANS-X Total Score

Mr CT scored 190 out of a possible 258 points on SPANS-X total score, substantially below the critical cut-off score of 227 for his age. This placed him in the <2nd percentile range, which furthermore the upper and lower limits of the 95% confidence interval also fell within the bounds of entirely. This "exceptionally low" result justified the further and more detailed exploration of SPANS-X index and subtest scores to formulate the cognitive deficits evident in his SPANS-X profile.

Table 1 Subtest base rates and percentiles

Subtest/Index	Raw score	Base rate	Percentile
Orientation to Person	4	99%	>1st
Orientation to Time	8	99%	>1st
Orientation to Place	4	98%	>2nd

Orientation to Condition	2	99%	>1st
Political Leadership	2	82%	>18th
Time Estimation	2	90%	>10th
(*Sum the 6 subtests above for*) **ORI**	22		
Digit Span Forward	5	31%	47th
Digit Span Backward	6	41%	>59th
Sustained and Divided I	10	87%	>13th
†Sustained and Divided II	8	13%	15th
†Counting Backwards	5	8%	35th
†Monetary Calculations	8	57%	>43rd
(*Sum the 6 subtests above for*) **ACI**	42		
Repetition	6	49%	>51st
Naming	10	0%	<2nd
Yes/No Questions	6	96%	>4th
Following Directions	6	87%	>13th
Reading	6	94%	>6th
Writing Sentences	5	71%	>29th
†Similarities	12	61%	>39th
(*Sum the 7 subtests above for*) **LAI**	51		
Object Recall	2	2%	2nd
Figures Recall	0	0%	<4th
List Learning	14	2%	20th
List Recall	1	0%	<4th
List Recognition	8	1%	1st
S-W P-A	3	0%	<1st
(*Sum the 6 subtests above for*) **MLI**	28		
Object Recognition	2	23%	24th
†Spatial Decision	7	3%	7th
Unusual Views	2	4%	4th
Figures Copy	9	0%	<4th
†Letter-Number Coding	7	6%	7th
Figures Recognition	1	4%	4th
Facial Expressions	3	2%	2nd
†3-and-1 Concept Test	16	79%	>21st
(*Sum the 8 subtests above for*) **VPI**	47		
†Sustained and Divided II	7	2%	2nd
†Spatial Decision	7	3%	7th
†Letter-Number Coding	7	6%	7th
†Counting Backwards	5	8%	35th

†Monetary Calculations	8	57%	>43rd
(*Sum the 5 subtests above for*) **ECI**	35		
†Similarities	12	61%	>39th
†3-and-1 Concept Test	16	79%	>21st
(*Sum the 2 subtests above for*) **CFI**	28		

† Score used in two SPANS-X indices.

Table 2 Raw index scores, index percentile (range) and critical value

Index	Raw score	Percentile (range)	C.O. (cut-off score)
ORI	22/22	50th	20.5
ACI	42/46	25th – 50th	39
LAI	51/53	50th	48
MLI	28/67	<2nd	54
VPI	47/70	<2nd	59
(Sum the 5 subtests above) SPANS-X TOTAL	190/258	<2nd	227
ECI	35/48	5th	39
CFI	28/28	75th	25

Table 3 Conversion of index score percentile range/confidence interval percentile range

Index	Raw score	95% C.I.	Raw score ± C.I.	C.I. percentile range
ORI	22	± 0.5	21.5 – 22	25th - 50th
ACI	42	± 2	40 - 44	10th – 75th
LAI	51	± 1	50 - 52	25th – 75th
MLI	28	± 3.5	24.5 – 31.5	<2nd - <2nd
VPI	47	± 3	44 - 50	<2nd – 5th
(Sum the 5 subtests above) SPANS-X TOTAL	190	± 7	183 - 197	<2nd - <2nd
ECI	35	± 2	33 - 37	2nd – 10th
CFI	28	± 1	27 - 28	50th – 75th

Orientation

Mr CT was able to answer all orientation questions correctly; however, this occurred in the context of providing several incorrect responses before he spontaneously corrected himself. He was aware of and named the hospital (having lived in the area for decades) in which the outpatient assessment appointment took place. The origin of his 'false starts' was unclear but knowing his history of becoming 'flustered' under pressure, it appeared to be an example of this. He appeared anxious and well-rehearsed, though stumbling over his lines. Every effort was made to calm him, ease his anxieties and establish good rapport. He earned points on the ORI for demonstrating some insight into his difficulties, though whatever his difficulties were, they did not yet have a name or label. Observationally, he provided more information than was necessary on some of his responses, for example providing a longer list of prime ministers than was requested, adding additional details about their policies and dates of term(s) served. This exhibited his extensive knowledge, yet appeared somewhat disinhibited, or an effort on his part to try to impress the administrator.

Attention/Concentration

Attention/concentration was a relative strength, scoring 42 of a possible 46 points on the ACI, reasonably above the critical cut-off score of 39. This score ranked him between the 25th and 50th percentiles, or "average", and 95% confidence interval between the 10th and 75th percentiles, "low average" to "high average". His single lower subtest performance, scoring at the 15th percentile, was on the Sustained and Divided Listening II subtest, which appeared to reflect difficulty multi-tasking or overloaded attentional capacity at moments while having to divide attention between competing demands. Observationally, Mr CT was vigilant, with good effort put to all tasks.

Language

Another of Mr CT's relative strengths was language, and he was very articulate in speech and sophisticated in content. His comprehension also appeared fine, and this observation was backed up by his subtest performances/ scores that required reasonable comprehension. He scored 51 out of a possible 53 points, above the critical cut-off score of 48, and placed him in the 50th percentile. The 95% confidence interval ranged from "average" to "high average".

Crucially, however, the Naming subtest was where he lost the two points on the LAI. He required a phonetic cue to name two of the six objects, often evident in 'word finding' difficulties. On the Naming subtest he scored <2nd percentile, with a base rate of 0%, meaning no one his age scored this low in the norming standardisation sample.

Memory/Learning

Mr CT's primary deficit was evident among the subtests that compose the MLI, scoring 28 out of a possible 67 points, substantially below the critical cut-off score of 54. This placed Mr CT's MLI entirely into the <2nd percentile range, including the 95% confidence interval range. The best subtest level performance was the List Learning subtest, where he scored at the 20th percentile, or "low average". This appeared to draw upon once again one of his greatest asset cognitive skills, that was his relative strength in immediate attention, span and working memory. Any distraction or delay from 'learning' or recall or recognition, however, resulted in significant loss of information.

He scored in the lowest possible percentiles among the remainder of MLI subtests, with base rates among the norming standardisation sample of 0% or only a small percentage (up to 2% of the standardisation sample). Importantly this was reflected in both delayed recall and recognition, very low recall and only essentially 'chance' recognition across the board, indicated significant and rapid loss of learning.

Visuo-motor performance

Mr CT scored 47 out of a possible 70 points on the VPI, substantially below the critical cut-off score of 59. This placed him squarely into the <2nd percentile, including the entire 95% confidence interval fell into this range, or 'exceptionally low' classification. Very few of the norming standardisation sample his age scored as low as he did, i.e., 0% to 6% on six of eight subtests. On the remaining two subtests he could have/seemingly scored 'chance' on one, and scored full points, >21st percentile on the 3-and-1 Concept Test.

Qualitatively, or interpreting Mr CT's lowest scores, visuo-spatial and visuo-constructional, hand-eye coordination skills seemed the most apparently changed from previous levels, where Mr CT would build models resembling science experiments, for example. On this testing occasion he seemed very left

hemisphere dominant in his style of approach to copying the Figure, constructing it piecemeal and from left-to-right, rather than relying on the Gestalt or the 'whole' of the Figure. On the Spatial Decision subtest he appeared to lack confidence in his perception of the dots and their location, and took a long time to respond, occasionally commenting what a challenge the test was, how the test had to be 'wrong'.

On the other hand, his perception and understanding/familiarity of 'objects' appeared relatively intact, albeit he did not always score full points on these subtests, dropping him once again into lower percentiles. In terms of function, however, it seemed Mr CT's visual difficulties were with 'where' and 'how' rather than 'what'. This conclusion seemed supported by other SPANS-X subtest performances, performing well on the more conceptual 3-and-1 Concept Test, knowing what objects were in the Naming subtest but just needing a phonetic (not semantic) cue to name them.

Efficiency

Mr CT scored 35 out of a possible 48 points on the ECI, below the critical cut-off score of 39. This placed him at the 5th percentile, or "below average". The 95% confidence interval placed him between the 2nd and 10th percentiles, or "below average" to "low average." This was interpreted as reduced efficiency in visuo-spatial and advanced attentional tasks, but not a global reduction in speed per se. Observationally, his response time, vigilance and the speed with which he undertook tasks, and his rate of speech were above average, but deficiencies in specific cognitive skills slowed him down and made him more prone to error – overall error, not speed, accounting for the reduced ECI score.

Conceptual flexibility

As may be expected of a man of Mr CT's intellect, he earned perfect scores with apparent ease on the CFI, overall scoring in the 'average' to 'high average' range. This suggested he could spontaneously form mental concepts from verbal and visual material, convert abstract concepts into superordinate categories, and produce alternative concepts (i.e. he demonstrated flexibility in thinking).

1.3. Conclusions

It appeared to be a strong probability that Mr CT was experiencing cognitive deterioration from his previously high-functioning levels. Particularly evident were rapid and nearly complete forgetting, complex or divided attention inefficiencies, naming/word-finding difficulties, and visuo-spatial/visuo-constructional impairments. These were evident both during psychometric testing and observation during two-plus hours of assessment and clinical interview and were consistent with some symptoms of Alzheimer's-type dementia, including the predominant memory impairment and deterioration from previous levels of everyday functioning. However, it also had to be noted that Mr CT retained many capabilities of independent living for the time being, including (most evidently to the assessment team) making it to every appointment at the hospital on his own, on time, and with all aspects of self-care and reasonably organised behaviour evident. On the other hand, Mr CT further demonstrated changes generally associated with the lateral and medial temporal lobes (i.e. learning and naming difficulties) and right parietal lobes (visuo-spatial/visuo-constructional and sustained attention), suggestive of simultaneous deteriorated functioning of specific regions of the brain.

The timing of having just moved did not seem opportune for Mr CT, changing his routine and living space. Given that he admitted feeling anxious, especially in terms of performance anxiety, it seemed appropriate to promote and arrange with Mr CT and his family his continued independence, with regular but non-intrusive

monitoring for as long as could be managed. Getting used to and organising his own space in his new living quarters was recommended as a useful first step, alongside someone with whom he felt comfortable and who could help him to get fully unpacked and organised. It was thought the changes and present state of chaos of his life added a psychological factor and layer of stress that Mr CT did not fully acknowledge.

This case seemed a clear organic deterioration of mental faculties, not entirely due to mood or psychological factors. Unfortunately, a diagnosis of early-onset Alzheimer's disease appeared probable, and thus earlier onset (i.e. aged 61) and more rapid deterioration was to be anticipated. His memory, including 'recognition memory' scores were too extremely low to be accounted for by psychological factors alone, and furthermore he subjectively denied feeling depressed, but rather anxious, which appeared related to decreased mental abilities and capacity to function, though both most probably fed into each other.

There did not seem to be an imminent need to make further drastic changes in Mr CT's life, but consideration of the possibility of further (perhaps even rapid) deterioration and getting the relevant affairs in order was advised to Mr CT and his family. He was required to report the probable diagnosis of early-onset Alzheimer's disease (which was later upheld by the neurology team/memory clinic) to the DVLA (driving and licensing agency) but again there did not seem an immediate need for him to stop driving, being very familiar with his neighbourhood, possessing vigilance and reasonable attention skills, and a spotless driving record to date. His assessment results, however, indicated that monitoring his driving abilities and preparing him for an inevitable loss of independence (at least for driving) was advisable, particularly in relation to the existing divided attention, visuo- spatial, and topographical orientation (and memory) difficulties. It was advised that he should be re-assessed within three months, using the same tests and the alternate version of SPANS-X (SPANS-X B) to both check the reliability of the current assessment findings, and/or to detect the trajectory of changes, and keep him and others safe.

1.4. Retest with SPANS-X B 3.5 months later

On this occasion Mr CT was accompanied by his daughter. It seemed the additional 3.5 months of disease progression, stress, and different assessment circumstances (i.e. having his daughter there), made Mr CT less engaging, and more anxious. Reports from his daughter indicated that "things had got worse", that for example, instead of getting unpacked and organised, Mr CT's new home had just become more unkempt and disorganised. Regarding his learning and memory, his daughter could not attest it was worse, but she did say he spent more time alone in his study, not functioning like his normal efficient and organised self and spending less time with the grandchildren.

Though probable early-onset Alzheimer's disease had already been diagnosed by the memory clinic team, a follow-up SPANS-X B retest seemed to support this diagnosis further, most notably he was now disoriented to the day of the week and the month, and he acknowledged less so the poor state of his condition on this occasion, more actively denying 'any problem'. His learning/memory, efficiency, complex attention, and visuo-spatial abilities were all significantly poorer as well.

Mr CT was officially advised to stop driving, and consider making arrangements for stepped memory care, from the totality of what turned out to be his final memory clinic visit. He was referred for an in-home assessment, with consideration of next steps for care.

Table 4 Critical difference (C.D.) for SPANS-X B retest

Index	SPANS-X B raw score	SPANS-X A raw score	B – A =	C.D.	0, +, or –
ORI	17	22	–5	1	-
ACI	40	42	–2	3	0
LAI	50	51	–1	1.5	0
MLI	21	28	–7	5	-
VPI	35	47	–12	4	-
(Sum the 5 subtests above) **SPANS-X TOTAL**	163	190	–27	10	-
ECI	28	35	–7	3	-
CFI	28	28	0	1.5	0

Key: 0 “no difference”; + “statistically significant improvement”; – “statistically significant decline”.