Françoise, a fifteen-year follow up

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Abstract – A fifteen-year follow up of the linguistic and cognitive profile of a woman, named Françoise, with standard trisomy 21. She had been studied in considerable detail between 1987 and 1991 by Rondal (1995) and found to have exceptionally good language abilities in advance of regular cognitive abilities for a person with Down syndrome. Françoise's psychological functioning has been deteriorating relatively rapidly over the last two years. A severe deterioration of her previously excellent receptive language abilities is documented in the present study. Productively, her on-line word finding is becoming problematic, mean length of utterance has halved compared with 15 years ago, and production of compound sentences is reduced. However, basic phonological and morphosyntactic skills are preserved. Françoise's changing profile during the latter years seems to mirror that documented in the first stages of Alzheimer disease in ageing adults in the general population.

Keywords: Down syndrome, Alzheimer disease, language

1. Background information

Between 1987 and 1991, one of us had the opportunity to analyse in detail the language and cognitive abilities of Françoise, a woman with standard trisomy 21, who had exceptionally good language abilities for a person with Down syndrome (cf. Rondal, 1995, for the full report). About two years ago, the day centre where Françoise spent several days a week, requested a neuropsychological examination because they were concerned about her depressed behaviour, memory losses, and spatial disorientation. M. Ylieff undertook the re-examination of some of Françoise's cognitive functions. Comparing his observations with those in Rondal's monograph (1995), Ylieff (2000) confirmed a marked deterioration of Françoise's episodic memory (Test of the 15 Words, Taylor's form A and B; Rey, 1964, 1967) and ability to deal with visuo-spatial and visuo-graphic material including a marked alteration of her writing and drawing performance with severe dysorthography. Pending subsequent neurological and neuroradiological examinations (which were refused by the family), Ylieff suggested that there was a pathology of the right hemisphere possibly linked to brain degeneration. No systematic evaluation of oral language was attempted. Based on informal conversations with Françoise, as well as on observations from the staff of the day centre, Ylieff's impression (Ylieff, May 2000, personal communication with J.A. Rondal) was that Françoise's overall productive language ability was mostly intact.

2. Follow-up study

In 2002, we repeated the analysis of Françoise's (now 47 years old) oral language ability. A deterioration in expressive language was found on this occasion, even on superficial contact, confirming the observations of the staff of the day centre which Françoise still attends one or two days a week. She spends the rest of the time in a residential facility for intellectually disabled adults, as her ageing mother (her father died in 1994) is unable to continue caring for her at home.

Table 1 summarizes a number of findings from the comparative assessment of Françoise's 2002 language abilities with that of 15 years ago.

As shown in Table 1, although Françoise's receptive lexicon has deteriorated it is still relatively preserved in comparison with other (i.e. nonexceptional) adults with Down syndrome (cf. Rondal, 1995; Rondal & Edwards, 1997). However, she experiences marked difficulties in finding words during conversation. Numerous dysfluencies characterise her spontaneous utterances compared to 15 years ago when she had none. Françoise's speech rate, judged previously to be normal, has slowed down considerably and her mean length of utterance (MLU) is half of its earlier value. She is keenly aware of her limitations in this respect and frequently expresses regret in not being able to find words as readily as she used to. However, Françoise's articulatory skills and basic productive morphosyntax have

TIME / FRANCOISE'S AGE		
LANGUAGE ABILITY	**1987-1991** **32-36 years**	**2002** **47 years**
1. Articulation 1.1. Segmental 1.2. Suprasegmental	 Fully correct Fully correct	 Correct in spontaneous speech. Hesitant prosody in longer utterances.
2. Dysfluencies	Virtually absent	Numerous speech dysfluencies (word and phrase repetitions, false starts, hesitations, undue pauses, broken utterances).
3. Approximate speech rate (in number of words per second)	3,3 words (12 to 15 phonemes)	1 word (4 to 5 phonemes) or less.
4. Receptive lexicon 4.1. TVAP 5-8[1] 4.2. TRT[2]	 28 correct designations out of 30 48 correct designations out of 50	 16 correct designations out of 30. 24 correct designations out of 50.
5. Productive morpho-syntax Mean length of utterance[3]	 12.34	 6.91
6. Receptive syntax 6.1. Relative clauses introduced by *qui* (who) 6.2. Causative subordinate clauses 6.3. Temporal subordinate clauses **6.4. Passive sentences** 6.4.1. Nonreversible 6.4.2. Reversible	 31/32 32/32 46/48 31/32 32/32	 1/16 5/32 0/16 5/32 7/32

Notes: 1. TVAP: Test de Vocabulaire Actif et Passif (Deltour, 1982); 2. Test des Relations Topologiques (Deltour & Hupkens, 1980); 3. Computed in number of words plus grammatical morphemes (Rondal, 1995).

Table 1. Comparative evaluation of Françoise's oral language abilities over time.

resisted the ongoing deterioration. In the three samples of conversational speech recorded, no articulatory error was detected nor any mistake in typical French word order. Nor did she exhibit any difficulty with the on-line production of obligatory grammatical morphemes marking gender, number, case, tense, or aspect. On the notoriously difficult test of the Non-word repetition (Borel-Maisonny, 1953), Françoise correctly repeated between 40% and 70% of the nonwords, presented orally and containing up to 4 syllables, as opposed to 100% correct repetitions fifteen years ago. However, the errors consisted of various perseverations and the omission, addition, or substitution of syllables, but not of sounds, suggesting attentional and short-term memory difficulties rather than articulatory limitations per se. Supporting this interpretation, on the balanced list of Rondal (1998), she could correctly repeat 72 out of 79 familiar words (mono-, bi-, and tri- syllabic ones); the seven errors all consisted of semantic confusions (e.g., *bras – arm* instead of *brave – brave*) or perseverations from the preceding item on the list.

In contrast stands Françoise's current receptive difficulties with the advanced syntactic structures that she had previously mastered. Her short-term memory span for digits is 2; for shorter phonologically similar words (containing one or two syllables) it is 1 (with perseverations); for shorter words phonologically dissimilar it is 2 (with perseverations); and for longer phonologically dissimilar words it is 1 (with perseverations). The corresponding 1989-1991 assessment yielded values of 4 digits, and 4 words or nonwords. Françoise's sentence span now averages about 4 words (with perseverations and substitutions of lexical items either semantically related or not related to the target elements). Fifteen years ago, Françoise's sentence span extended to 14 words. She has markedly increased attentional difficulties. On the Barrage Subtest of the KLT Scale (Kettler, Laurent, & Thireau, 1964), Françoise is now only able to deal (unsystematically) with a few stimuli, while she scored 22 out of 90 (lowest quartile of the typical population) 11 years ago.

Waters, Caplan and Rochon (1995) have observed that patients with probable[1] Alzheimer disease, with no previous

[1.] *Probable* Alzheimer disease because the condition can only be certified upon post-mortem brain anatomical examination. Peripheral biological markers and neuroimaging can be used, however, as predictive indications in the general population and also in persons with Down syndrome (cf. Percy, 1993, and Schapiro, 1993).

impairments, have difficulties that are not so much concerned with the syntactic structures themselves, but rather with assigning sentence meaning and using it to search for the truth of a given meaning in semantic memory. This suggests that the syntactic impairment of sentence comprehension found in probable Alzheimer patients by some researchers (e.g. Emery, 1988), but not by others (e.g. Schwartz, Marin & Saffran, 1979; Appel, Kertesz & Fishman, 1982; Rochon, Waters & Caplan, 1994; Ruiz, 1998), could be due to a disruption in the ability to use sentence meaning to accomplish post-interpretive tasks.

In the case of Françoise, the possibility cannot be ruled out, on the basis of the preceding examination, that she could have retained at least part of her receptive syntactic competence but is experiencing serious performance difficulties either in establishing lexical meaning within sentences, keeping lexical meaning in short-term memory for deriving sentence interpretation, and/or accomplishing post-interpretive tasks. With this possibility in mind, we have re-tested Françoise on a series of 16 affirmative full passive sentences (plausibly reversible and non-plausibly reversible), constructed around actional (8), experiential (4), and affective verbs (4). These sentences could be regarded as having simplified non-structural features in comparison to the set of sentences used previously, as the vocabulary items involved were of a higher level of frequency in the French language. The sentences were read slowly one by one to Françoise by the examiner. Two questions were then asked orally. **Question one:** "Who is Xed ?" (where X stands for the verb in the sentence to be interpreted). **Question two:** "Who is Xed by Y ?" (where Y stands for the semantic agent expressed as oblique object in the surface sentence). Table 2 lists the 16 sentences together with Françoise's responses.

As shown in Table 2, Françoise gave only two correct answers to Question one (12.5%; i.e. much below chance level, in this case 50%) and no correct answers to Question two, in spite of the fact that the answer to the latter question was extremely easy as it related in all cases to the first noun phrase. Françoise seemed often to be led astray by incorrect lexical associations. For example, in sentence 3, having identified the past participle *mordu/e* (bitten) of the verb *mordre* (bite), she ignored the rest of the sentence, gave the response *chien (dog)*, and repeated this in sentence 4. A similar situation prevailed in sentences 5 to 12 (initially with the pair *soigner [to look after]; médecin [physician]*) as well as in sentence 14 to 16 (with the pair *aimer [love]; garçon [boy]*). An interesting phenomenon happened in sentences 5, and again in sentence 14, revealing how slow Françoise's language processing has become. In sentence 5, following her incorrect answer to question two, she added: *C'est le médecin qui est mordu par le chien (It is the physician who is bitten by the dog)*, demonstrating that she was still partly processing the verbal content of sentences 3 and 4, and producing for that matter, a syntactically correct pseudorelative in the passive voice. Similarly, asked about sentence 13 *(Le garçon est aimé par la fille – The boy is loved by the girl)*, she declared on two successive occasions that she did not know the answer to the question. However, in

answering the questions following sentence 14, she used the word *garçon (boy)*, which had not appeared in sentence 14 but was present in sentence 13, and persevered with it in sentences 15 and 16.

Pursuing our attempt at disentangling Françoise's linguistic competence and performance limitations, we presented four full passive sentences (two plausible and plausibly reversible, and two plausible but not plausibly reversible ones) taken from the test E.CO.S.SE (Lecocq, 1996). Françoise was requested to point towards one picture in an array of four in response to the same two questions as before. This reduced the demand of the interpretive task in several ways (e.g. the need to keep in mind the referents of the nominal and verbal elements for receptive analysis, and the need to produce a verbal response to the examiner's questions are removed). Table 3 lists the sentences used in this part of the study together with the non-verbal responses supplied by Françoise.

As shown in Table 3, Françoise correctly responded two times in four to Question one, that is at chance level, and no times in four to Question two (with a lexical perseveration from sentence 2 to sentence 3).

Unfortunately, testing could not be pursued any further as the medical staff of the centre intimated that given Françoise's mental state of mind and her difficulties with the tasks, it was not advisable to continue with the examination.

A check on the latest sample of spontaneous speech collected from Françoise confirms that despite the slowness of her productions and the numerous dysfluencies, her MLU is still close to 6 units, the canonical word order is correct, and the obligatory grammatical morphemes correctly produced without hesitation. Phoneme coarticulation in spontaneous speech is correct even if prosody over longer utterances tends to be hesitant. Despite her receptive difficulties with complex syntactic structures, Françoise is still able to produce a few two-clause sentences [mainly completive and pseudorelative ones; e.g., *Et puis alors, on attend que les autres soient là (And then, one waits till the others are there); C'est l'éducatrice qui le fait elle-même (It is the educator who does it herself)*].

Françoise's deterioration suggests that an excellent level language achieved by a person with Down syndrome is no guarantee against later deterioration. This contradicts our previous hypothesis, i.e., the more developed a psychological function, the better it should resist brain degeneration (Rondal & Comblain, 2002). Either such an opinion is false or it does not apply to individuals with Down syndrome.

3. Language difficulties in Alzheimer disease

Progressive difficulty in retrieving words from long-term memory in naming referents and constructing sentences is one of the earliest symptoms of Alzheimer disease (e.g. Bayles & Tomoeda, 1983; Flicker, Ferris, Crook & Bartus,

SENTENCES	RESPONSES
I. La fille est mordue par le garçon (The girl is bitten by the boy)	I. le chien (the dog) 2. le chien (the dog)
2. Le livre est jeté par le garçon (The book is thrown away by the boy)	I. le livre (the book) 2. Françoise says : "Il ne veut plus jouer avec des jeux." (He does not want to play games anymore).
3. Le garçon est mordu par le coussin (The boy is bitten by the cushion)	I. le chien (the dog) 2. le chien (the dog)
4. La table est mordue par le livre (The table is bitten by the book)	I. le chien (the dog) 2. le chien (the dog)
5. La fille est soignée par le garçon (The girl is looked after by the boy)	I. le médecin (the physician) 2. le médecin (the physician) Françoise adds: "C'est le médecin qui est mordu par le chien." (It is the physician who is bitten by the dog).
6. L'oiseau est soigné par la fille (The bird is looked after by the girl)	I. le médecin (the physician) 2. le médecin (the physician)
7. La fille est soignée par l'oiseau (The girl is looked after by the bird)	I. le médecin (the physician) 2. le médecin (the physician
8. Le livre est soigné par la table (The book is looked after by the table)	I. le médecin (the physician) 2. le médecin (the physician)
9. La fille est vue par le garçon (The girl is seen by the boy)	I. le médecin (the physician) 2. le médecin (the physician)
10. La boîte est vue par le garçon (The box is seen by the boy)	I. le médecin (the physician) 2. le médecin (the physician)
II. Le garçon est vu par la boîte (The boy is seen by the box)	I. le médecin (the physician) 2. le médecin (the physician)
12. La télévision est vue par la boîte (The TV set is seen by the box)	I. le médecin (the physician) 2. le médecin (the physician)
13. Le garçon est aimé par la fille (The boy is loved by the girl)	I. Françoise says that she does not know 2. Idem
14. Le livre est aimé par la fille (The book is loved by the girl)	I. le garçon (the boy) 2. le garçon (the boy)
15. La fille est aimée par le livre (The girl is loved by the book)	I. le garçon (the boy) 2. le garçon (the boy)
16. Le vélo est aimé par le livre (The bike is loved by the book)	I. le vélo (the bike) 2. le garçon (the boy)

Table 2. Series of 16 lexically simple plausible and implausible, reversible and non reversible, affirmative passive sentences given to Françoise in the order and Françoise's responses to the two interpretive questions (1. "Who is Xed ?"; 2. "Who is Xed by Y ?").

1987; Murdoch, Chenery, Wilks & Boyle, 1987). This lexical difficulty primarily involves word production and not comprehension. The early deterioration in word finding in Alzheimer disease patients is supported by two kinds of evidence: (1) lower scores on standard tests of naming; and (2) spontaneous speech marked by circumlocution, over reliance on empty forms of reference (e.g., pronouns or vague content words like "thing" or "guy" and word-finding episodes terminating with failure comments such as "I can't remember that one"). Neuropathological aging is also commonly characterized by word retrieval difficulties (related to frequency and recency of word use; Burke & Laver, 1990). With regard to individuals with Down syndrome, Kledaras, McIlvane and MacKay (1989) have reported a decline in the ability to name pictures in a 59-year-old man (JB) with standard trisomy 21 who was diagnosed as having a dementing illness of the Alzheimer type. Over the last 10 months of observation, his naming scores

SENTENCES	RESPONSES
1. La fille est poussée par le cheval (The girl is pushed by the horse)	1. le cheval (the horse) 2. Françoise says that she does not know
2. Le camion est poussé par le garçon (The truck is pushed by the boy)	1. le garçon (the boy) 2. Françoise says that she does not know
3. Le cheval est poursuivi par l'homme (The horse is run after by the gentleman)	1. le cheval (the horse) 2. le garçon (the boy)
4. Le landau est poussé par le monsieur (The landau is pushed by the gentleman)	1. le landau (the landau) 2. Françoise says that she does not know

Table 3. Sentences from the E. CO.S.SE battery and Françoise's picture designations in response to the two interpretive questions (1. "Who is Xed"; 2. "Who is Xed by Y).

on a list of 40 common lexical items selected by the authors, declined gradually at first and then precipitously until they were less than 30% correct on the final test. Error analysis showed that lower scores generally reflected loss of stimulus control by relevant characteristics of the test pictures and a shift towards irrelevant features of the task. Early on, JB's naming errors belonged to the same semantic category as the target stimulus. Later, his errors became more indiscriminate and perseverative reflecting the increasing influence of irrelevant stimuli. A corresponding pattern of deterioration has been observed in naming tasks in typically developed adults with Alzheimer disease by Schwartz et al. (1979) and Flicker et al. (1987).

As mentioned above, several researchers have suggested that there may be little genuine syntactic impairment in the early stages of Alzheimer disease in typically developed adults. Françoise has retained a minimal productive ability consisting of morphosyntactic sequences of six words or so, correctly articulated and meaningfully related even if on-line word finding has become difficult with a high frequency of dysfluencies. Her receptive treatment of multiword utterances is more problematic. The test results indicate serious impairments as she now demonstrates: sentence span lower than MLU, numerous perseverations and substitutions of lexical items, and major attentional difficulties. Françoise appears to have lost most of her ability to deal with complex syntactic structures receptively. However, the possibility, as remote as it may seem, that she has kept at least a part of this complex syntactic ability but that the massive performance limitations she is facing now prevent her from sharing in interpretive tasks and sustaining longer productions, cannot be completely ruled out.

Difficulties in sentence comprehension in typically developed persons with dementia of the Alzheimer type have been attributed to impairments in some of the mechanisms involved in verbal short-term memory, such as storage of phonological forms and rehearsal (e.g. Just & Carpenter, 1992). Such a causal implication appears to be based on a line of research stressing the reliance of sentence comprehension upon short-term memory capacity (MacDonald,

Just & Carpenter, 1992; see Just & Carpenter, 1992, for a review) and on the observation that short-term memory functioning (span, rehearsal, retrieval, and processing speed) is negatively correlated with age in elderly adults (e.g. Gerard, Zacks, Hasher & Ravansky, 1991; Norman, Kemper & Kynette, 1992). This view has been challenged on empirical grounds, in nondemented individuals and patients with probable Alzheimer disease. For instance, it has been shown (cf. Waters et al., 1995) that typically developed adults with varying short-term memory spans do not differ in the comprehension of particular syntactic structures. Correspondingly, some patients with reduced short-term memory spans have been shown to be capable of excellent sentence comprehension, including normal comprehension of sentences containing syntactically complex forms (e.g. Waters & Caplan, 1996, 1997).[2] In the same way, Almor, MacDonald, Kempler, Andersen and Tyler (2001) have observed that, neither in memory impaired patients with probable Alzheimer disease nor in healthy elderly control participants, is performance on subject-verb number agreement in short sentences correlated with performance on short-term memory tasks. This suggests that the processes involved in interpreting the grammatical dependency between adjacent and nonadjacent elements in sentences are different from those required in short-term memory tasks.[3] What may be involved instead is the common observation that individuals with probable Alzheimer disease have impairments of the central executive component in the working memory system proposed by Baddeley (1980). Such patients have been shown to be impaired in tasks involving attention shifting and other executive functions (e.g. Morris, 1986; Nebes & Brady, 1989; Baddeley, Bressi, Della Sala, Logie & Spinnler, 1991).

Regarding sentence production, typically developed patients with Alzheimer type diseases have been reported to exhibit preserved syntactic ability in the contexts of impaired semantic ability (e.g. Blanken, Dittman, Haas & Wallesch, 1987; Illes, 1989). Kempler, Curtiss and Jackson (1987) have assessed the spontaneous speech of twenty

[2.] For empirically based suggestions as to a limited role of short-term memory in morphosyntactic functioning, see, for example, Butterworth, Campbell and Howard (1986), Butterworth, Shallice and Watson (1990), and Rondal (1995).
[3.] Almor et al.'s data (2001) also show that the short-term memory deficits of typically developed adults with probable Alzheimer disease do interfere with the discourse processing demands, i.e. the participants' on-line treatment of the content dependencies across sentences.

patients with probable Alzheimer disease. The analysis revealed a typical range and frequency of correctly formulated syntactic constructions (i.e. simple and conjoined sentences, questions, relative, adverbial, and infinitival clause, passives) but poor lexical use. Bates, Marchman, Harris and Wulfeck (1993), however, have questioned the suggestion that grammatical production is unimpaired in the first stages of Alzheimer disease. They have reported data purportedly showing that when grammar is assessed under highly constrained conditions (e.g. in a film description task), production is impaired in the sense that these patients have difficulties in accessing adequate fits between meaning and combinatorial forms due to limited processing resources. However, upon close examination, Bates et al.'s data turn out to be compatible with the above data concerning the preservation of basic productive morphosyntactic ability in probable Alzheimer disease patients. Bates et al. have demonstrated with seven females and nine males diagnosed with probable Alzheimer disease (aged between 65 and 92 years), that there is indeed a reduction in the production of complex syntactic forms, e.g. full passives, multi-clause sentences, and plurisentences, in describing two conjoined or sequential events. However, the range of complex structures that they use is not different from healthy controls or younger subjects. If the processing resources available to patients with probable Alzheimer disease are indeed reduced (cognitively and semantically), it does not seem that profound or even marked differences in productive morphosyntax at this stage are the rule between these patients and nonpathological ageing people. Moreover, Bates et al. have not ruled out the possibility that the reduction of complex syntactic forms in patients with probable Alzheimer disease could be better attributed to their difficulties with on-line word finding in sentence elaboration and subsequent diminished risk taking.

In contrast to sentence production, there is a consensus that discursive functioning is usually severely altered in Alzheimer disease. Arkin and Mahendra (2001) have reviewed a dozen discourse studies documenting decreased cohesion, more use of egocentric and exophoric references, frequent omissions of referents, poor topic maintenance, ideational perseveration, and difficulties with the macrostructures of the various types of discourse (e.g. narrative, argumentative) in persons with probable Alzheimer disease.

However, it is essential to consider the evolution of the signs and symptoms in Alzheimer disease. Ruiz (1998) has analyzed the outcomes of several longitudinal studies of the condition in typically developed ageing adults. Briefly summarized, in the *first stage* (with an average duration between 2 and 4 years), one may observe a diminution of the expressive ability and gradually more marked difficulties in retrieving lexical items from semantic memory. At this stage, comprehension deficits are not typical. In the *second stage*, the expressive deficit is more marked in conversational speech, with repetitions and an inability to detect them, and problems of on-line comprehension appear. At this second stage, it would seem that the morphosyntactic

ability (particularly on the expressive side) is not typically affected. In the *third stage*, the degeneration of the brain is more advanced and there is more general deterioration of neuropsychological functioning. Behavioural and communicative difficulties are more marked with a most noticeable reduction of spontaneous speech. When language is produced, it is characterized by numerous lexical and semantical errors, echolalia, anomia, and phonemic transpositions. Morphosyntactic limitations are evident. Problems with sentence comprehension become more serious than before, with difficulties in understanding even simple verbal orders. At this third stage of the condition, serious alterations of the pragmatic aspects of language are taking place. In some cases, the patients fail to respond to the questions asked, or supply answers that have little or no relationship with the semantic contents of the questions. In the *fourth and last stage*, which usually precedes death by approximately one year (Caballero, 1991), the clinical picture is that of global aphasia with mutism.

4. Linguistic and cognitive ageing in Down syndrome

Before interpreting Françoise's cognitive and language evolution over the latter years, it is necessary to take into account a possible "natural" language and cognition deterioration with ageing that might occur in people with Down syndrome after 40 years of age and who do not have a specific neurological disorder such as Alzheimer's (Brown, 1985). The likelihood of an ageing process occurring earlier in Down syndrome than in other syndromes of intellectual disability has been suggested (e.g. Thompson, 1999). It would seem that physically and biochemically some earlier ageing process may be at work in Down syndrome already identifiable in the forties and possibly before (Van Buggenhout, Lukusa, Trommelen, De Bal, Hamel & Fryns, 2001; Franceschi, 1998). Cognitively and linguistically, however, the issues are less clear. Fenner, Hewitt and Torpy (1987) have reported a decline in mental age in less than one-third of their total sample (n=39) of persons with Down syndrome between 20 and 49 years and in just over one-third of those older than 35 years. Ribes and Sanny (2000) have documented a reduction in short-term and long-term memory, in vocabulary use, and in expressive as well receptive language abilities in adults with Down syndrome. According to their data, there is already a slight decline in cognitive and language abilities evaluated between 20 and 40 years. However, a more marked decline takes place beyond 40 years. Along similar lines, Moss, Tomoeda and Bayles (2000) have reported a significant inverse relation between increasing age and several aspects of auditory linguistic comprehension in a cohort of individuals with Down syndrome aged between 32 and 65 years. Correspondingly, Prasher (1996) has documented an age-associated decline in short-term memory, speech, practical skills, general level of activity and interests, in approximately 20% of the persons with Down syndrome aged 50 to 71 years.

But other researchers are less definitive on the topic. Little to no change in nonverbal reasoning, memory, language (receptive and expressive vocabulary), planning and attention, perceptual-motor and adaptive skills until close to sixty years is suggested in a study by Das, Divis, Alexander, Parrila and Naglieri (1995). The same authors indicate, however, that the older people with Down syndrome in their cohort (i.e. those over sixty years) showed poorer performances than those in younger groups, particularly on tasks requiring attention and planning. George, Thewis, Van der Linden, Salmon and Rondal (2001) have conducted a four-year longitudinal study of 12 participants with Down syndrome (six women and six men), aged between 36 and 48 years at the beginning of the study (cf. George, Thewis, Van der Linden, Salmon & Rondal, 2001; Rondal, in press). Their language functions (receptive as well as productive; with tasks concerning the lexical, morphosyntactic, and discursive aspects of language) were assessed at one year intervals as well as a number of nonverbal cognitive abilities [auditory-verbal and visuo-spatial short-term memory, episodic memory; using an adaptation of the child Rivermead Behavioural Memory Test (Wilson, Ivani-Chalian & Aldrich, 1991); visual perception, visuo-spatial functions, executive functions, reasoning (evaluated with the K-ABC, Kaufman & Kaufman, 1993), and attention]. Four participants did not maintain their involvement beyond the second year of study. None of the analyses yielded a significant result, failing to corroborate the null hypothesis of a language change and/or a change in nonverbal cognitive functions across and over the four years of study. Comparing that part of the above language data obtained with the use of the receptive subtests of the Batterie pour l'Evaluation de la Morpho-Syntaxe (Comblain, 1995) with corresponding data reported by Comblain (1996) from her study of adolescents (mean chronological age: 16 years and 7 months) and younger adults (mean chronological age: 26 years and 9 months).[4] Rondal and Comblain (2002) showed that no marked change takes place in receptive morphosyntactic abilities of persons with Down syndrome in the time between late adolescence and roughly fifty years of age.

Other longitudinal studies have contributed observations with similar results. Devenny, Hill, Patxot, Silverman and Wisniewski (1992), and Burt, Loveland, Chen, Chuang, Lewis and Cherry (1995) did not observe significant changes in the cognitive functioning of individuals with Down syndrome aged between 27 and 55 years, and 22 and 56 years, in the two studies respectively, over intervals from 3 to 5 years. Devenny, Silverman, Hill, Jenkins, Sersen and Wisniewski (1996) report only four cases of cognitive decline in 91 adults with Down syndrome followed for five years beyond the age of fifty.

5. Interpreting Françoise's linguistic and cognitive evolution

From the above indications, it seems clear that Françoise's change over the latter two years is specific. It cannot be attributed to a gradual loss of her language and cognition abilities over time since such a decline is not obvious at least until fifty years of age in persons with Down syndrome. Françoise's recent evolution seems to correspond rather closely to that seen in typically developed persons with probable Alzheimer disease. At the present stage, she may be a probable Alzheimer disease patient. It is known that this pathological condition, apart from any effect of possible earlier ageing, affects between 25 and 45% of persons with Down syndrome beyond 55 years (Dalton & Crapper-McLachlan, 1984; Lai & Williams, 1989; Zigman, Schup, Haaveman & Silverman, 1997; Wisniewski & Silverman 1999). It is less common before that age but no clear statistics are available.

Comparing Françoise's recent language and cognitive evolution with the evolution of the language in typically developed ageing adults with Alzheimer disease (Ruiz, 1998), one cannot but note the close correspondence between the two, with the additional indication that Françoise's deterioration has been more rapid than is usually the case in typically developed adults.[5] Another case of probable Alzheimer disease in a woman with Down syndrome with degeneration over a period of three years from the age of 43 has been reported by Tyler and Shank (1996), without language assessment but with cranial axial computerized tomographic images confirming atrophic changes in her brain. She has the symptoms of urinary incontinence, tonic-clonic seizures, dehydration due to her refusal to swallow liquids as well as solid food, and delirium. This may argue against the not uncommon suggestion that for those individuals with Down syndrome who develop Alzheimer disease, there may be a ten-year latency or more between the presence of critical Alzheimer disease-type neuropathological changes and clinical signs and symptoms, as opposed to 4 or 5 years in the typical population (Wisniewski, Dalton & Crapper-McLachlan, 1985; Lai & Williams, 1989; Wisniewski & Silverman, 1996).

At the present time, Françoise seems to have reached the third stage identified by Ruiz (1998), i.e. noticeable reduction of spontaneous speech, numerous lexical and semantical errors with anomia and perseveration, and morphosyntactic shortcomings, still limited in expressive

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[4.] The three cohorts having comparable mental age; (adolescents: 4 years and 4 months, standard deviation: 8 months; younger adults: 4 years and 7 months, standard deviation: 9 months; older adults: 4 years and 4 months, standard deviation: 6 months.
[5.] Such a comparison is meaningful only because Françoise's point of departure in language ability, so to speak, is close to normal (see also Campbell-Taylor, 1993), as attested by the investigation carried fifteen years age by Rondal (1995). For typical persons with Down syndrome, it has been reported (e.g. Moss et al., 2000) that in terms of visuospatial construction and language comprehension, nondemented adults with Down syndrome perform more poorly than typically developed individuals with mild and moderate Alzheimer's disease (corresponding to Ruiz' first two stages).

language, but marked in sentence and discourse comprehension, including even with simple clauses or utterances. Pragmatically, however, it is not clear that Françoise conforms to Ruiz's stage three. She still has a good sense of social interaction, verbally and nonverbally. Whenever she fails to respond adequately to a question or to give the proper social answer, this seems to be due more to her semantic and lexical receptive difficulties than to an apparent breakdown of pragmatic competence.

Françoise, like typically developed ageing adults with probable Alzheimer disease, has retained at this point most of her co-articulatory and basic expressive morphosyntactic abilities. It is relevant to see whether the explanations advanced in the Alzheimer's literature in order to explain this type of dissociation between language components, apply to Françoise's case.

Ruiz (1998) suggests that semantic-pragmatic processing in language functioning requires "more complex operations" which may rely particularly on memory processes and other mental systems. This is a curious argument given that the generative school in linguistics (e.g. various Chomsky's writings and opinions, cf., in particular, Rondal, 1994) has always sustained that syntax and phonology are more complex. Actually, we have no suitable measures to enable us to compare the relative complexity of the various components of the language system, which probably all have their own particular type of complexity. The complexity argument, therefore, is not a good one.

Kempler et al. (1987) claim that such observations as the above mentioned dissociation in the language organization of probable Alzheimer disease patients are consistent with a modular theory of language (e.g. Chomsky, 1981). More precisely, they suggest that the overlearned and more automatic nature of syntactic ability can help account for its relative resilience to some degree of cognitive loss and cortical degeneration. Automatic processes operate independently of the individual's control, do not require attention, and do not use short-term memory capacity (Schneider & Shiffrin, 1977). Regarding language functioning, automatic processes appear to develop when the range of alternatives is well defined and limited (as is the case for phonology and morphosyntax) as opposed to components where the range of alternatives is broad and much less or not at all predictable (as is the case for semantic and discursive events). In the latter cases, more 'on-line' processes are usually called upon. Of course, phonology and morphosyntax are not automatic in some absolute sense, but they are markedly more automatic, once acquired than such processes as lexical selection and discourse organization. Automaticity has

been used to explain why certain skills once acquired are maintained in the case of brain damage (e.g. Jorm, 1986). If phonology and basic morphosyntax are largely automatic in the adults, and automatic processes tend to be preserved in brain damage and in the early phases of brain degeneration, one should observe maintenance of those productive aspects of language in probable Alzheimer disease patients including those with Down syndrome, to the extent that the latter have developed sufficiently in these respects. And this is indeed what is observed with typically developed adults with probable Alzheimer disease as well as in Françoise's latest language production. Automatic combinatory sequences of words and inflectional morphemes are maintained whereas lexical selection, further clause and sentence organization, and discourse regulation, which require controlled processes, are affected from the early stages of the pathological evolution.[6]

Another way of explaining these phenomena is the theory that different aspects of language functioning are based on different memory systems. For example, language comprehension presupposes the concurrent activation of semantic (explicit) memory, which is responsible for lexical recognition, and of procedural (implicit) memory,[7] which is responsible for grammatical comprehension (Fabbro, 1999). Explicit memory is conscious by definition, whereas implicit memory is not. Based on neuroimaging studies, it is known that during semantic memory tasks, the prefrontal, the cingulate (association fibers connecting the frontal and temporal lobes) and the superior temporal cortex of the left-cerebral hemisphere are activated (Squire & Zola-Morgan, 1991). During procedural memory tasks, prefrontal areas of both cerebral hemispheres, the basal ganglia and the cerebellar hemispheres are activated (Salmon & Butters, 1995). The automatic processes intervening in simple combinatorial language production, referred to by Kempler et al. (1987), depend on procedural memory whereas lexical selection and recognition as well as further discourse organization and regulation, as more controlled processes, are more dependent on semantic memory. It is likely that in Françoise's case, as well as in the case of typically developed stage-three Alzheimer patients (in Ruiz' taxonomical scheme), there is a marked degradation of the prefrontal, cingulate, and superior temporal cortical areas responsible for the major problems encountered regarding language comprehension and complex structural functioning [in addition to damages to other prefrontal areas of both cerebral hemispheres, as well as to the left superior temporal and the right parietal ones, typically considered to be devoted short-term memory territories (cf. Meunier, Bachevalier & Mishkin, 1994)] whereas the basal ganglia

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[6.] Interestingly, Kempler et al. (1987) remark that there is a parallel here with the preservation of overlearned motor function in the first stages of the evolution in Alzheimer disease. Fully automatic motor processes, such as walking or smoking, are preserved. It is when new information needs to be integrated or a detour conduct initiated to overcome a particular obstacle that deficits appear. This may be extended to speech articulation.
[7.] According to current classifications, long-term memory consists of explicit memory, which is divided into semantic (the system storing the subject's knowledge of the world and the meaning of the words) and episodic or autobiographical memory, and of implicit memory, which contains procedural memory (a type of knowledge depending on the repeated execution of a task, even though the subject may not be aware of the nature and extension of the implicit knowledge s/he has actually stored), and other components (i.e. priming and conditioning memory which are of no direct interest here) (cf. Tulving, 1987, and Schacter, 1996).

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and the cerebellar hemispheres are preserved to a large extent and the bilateral nature of the prefrontal participation at this level of functioning allows for a preservation of automatic procedures despite damages to some of the latter areas.

Conclusion

An important aspect of the above discussion is that the description of Françoise's most recent language and cognitive changes conform rather well to the changes reported in the literature on probable and confirmed Alzheimer disease in typically developed ageing adults, except for the increasingly rapid speed of functional degeneration in Françoise's skills over the last months. A clear dissociation is observed in Françoise's contemporary language functioning between comprehension and production of speech sequences, and, within language production, between the semantic and informational aspects of language (badly damaged) and the basic phonological and morphosyntactic ones (still relatively preserved). These two dissociations (particularly the second one) were more easily and clearly identifiable in Françoise given that her superior linguistic talents (both receptive and expressive) for a person with Down syndrome had been carefully documented previously.

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