Down Syndrome Research and Practice Vol 5, No.1, pp 1 - 15

ISSN: 0968-7912 (1998) 5:1

CASES OF EXCEPTIONAL LANGUAGE IN MENTAL RETARDATION AND DOWN SYNDROME: EXPLANATORY PERSPECTIVES

Jean A. Rondal

(From a keynote presentation to the 2nd International Conference on Language and Cognitive Development in Down syndrome, Portsmouth, UK, April 8-10, 1997.)

Jean A. RONDAL, Ph.D., Dr. Ling.

Recent studies of exceptional language development and functioning in mentally retarded people raise questions regarding basic issues in language disorders. These studies are summarised and their implications discussed. Possible reasons for the existence of such cases are examined including language training, general cognitive functioning, working memory, cerebral dominance, and deep-seated variation at brain level.

Keywords: Down syndrome, language development, exceptional language development, brain development

1. Exceptional language in the present context refers to levels of functioning not usually found in moderately and severely mentally retarded (MR) persons although they are the norms for nonretarded (NR) people.

Table 1 summarises a series of cases of exceptional language development appeared in the literature.

Ursula Bellugi (1997) points out that the three cases that she documented in her 1988 paper (with Marks, Bihrle, & Sabo) may be standard for Williams syndrome (WS) adolescents as judged from recent studies (Singer et al., unpublished; Karmiloff-Smith et al., 1995, 1997; although it cannot be said that the morphosyntax of WS individuals is intact). Case no. 5 (Françoise) - Rondal, 1995 - is detailed in Table 2. The study of case no. 9 (Claudine) - Table 3 - is still in progress. Claudine's receptive language abilities have not been systematically evaluated yet.

The conclusion to the above studies is straightforward. Formal language problems are not inherent in MR/Down syndrome (DS) qua MR/DS. I suggest that the problems of MR/DS persons in the semantic aspects of language, on the one hand, and in the phonological and morphosyntactic aspects, on the other, do not have the same roots. The semantic problems of MR/DS subjects with language originate in their cognitive difficulties. These problems are unavoidable given that major cognitive limitations define the MR condition. On the contrary, the difficulties of typical MR/DS subjects with the formal aspects of language (at least the advanced ones) do not originate in general cognition, as demonstrated by the exceptional cases. My opinion is that they result from specific impairments in language organisation. This runs contrary to the view that all language difficulties in MR are a direct consequence of their cognitive deficit.

2. The exceptional cases of language development and functioning identified demand an explanation. I will undertake this risky exercise relying particularly on the Françoise case, because this is the case for which I have the most complete record. I will quote from other exceptional cases whenever relevant information is available. Four

Table 1- Exceptional cases of language development and functioning in MR/DS subjects. Studies and keypoints.

Study	Subjects	CA[1]	IQ[2]	Operational level[3]	MA[4]	MLU[5]	Other Language aspects
1. Bellugi, Marks, Bihrle & Sabo (1988)	Van Crystal Ben (Williams syndrome)	11 15 16	50 49 54	Preoperatory Preoperatory Preoperatory		8.60 13.10 10.00	- Correct articulation and phoneme discrimination - Receptive vocabulary at the 9-12 year level level - Advanced expressive and receptive morphosyntactic abilities
2. (Curtiss and associates 1989, Yamada, 1990)	Antony Rick Laura (etiologies unknown)	6 15 16	50 41	Preoperatory Preoperatory Preoperaory	2 years 9 mths		- Correct articulation and phoneme discrimination - Receptive vocabulary at the 6 year level and lower. - Advanced expressive and receptive morphosyntactic abilities, except for Laura who exhibited receptive morphosyntactic limitations. - Semantic pragmatic and discursive deficiencies
3. Seagoe (1965)	Paul (Down syndrome)	11	60				- Good command of written language expression and reading (average number of words per written sentence varying from 7.14 to 12.50 between 15 and 33 years).
4. Hadenius, Hagberg, Hyttnas-Bensch, & Sjogren (1962), Anderson & Spain, (1977) Tew (1979)	Hydrocephalic children	3	Severely impaired cognitive development				- Good ability to articulate, learn words, and use complex syntax - Semantic deficiencies
5. Rondal (1994a, 19994b)	Françoise (Down syndrome)	32	60	Late preoperatory to early operatory	7 years 4 mths	12.24	- Correct articulation and phoneme discrimination - Moderately retarded lexical development - Advanced expressive and receptive morphosyntax - Limitations in discourse organization
6. Cromer (1991)	DH (etiology unknown)	Adoles -cent	35				- Correct articulation - Extensive vocabulary - Complex morphosyntax - Use of standard pragmatic devices in conversation.
7. O'Connor & Hermelin (1991) Smith & Tsimpli (1995)	Christopher (etiology unknown)	29	67	Preoperatory			- Practice of English within the normal range including complex metalinguistic judgements. - Good level ability in translating in English from 13 languages: French, German, Spanish, Danish, Dutch, Finnish, Russian, Greek, Hindi, Norwegian, Polish, Portuguese, and Welsh.
8. Vallar & Papagno (1993)	FF (Down syndrome)	23	71				- Correct articulation with occasional suttering like phenomena - Good acquisition of Italian and to a lesser degree of English and French vocabularies. - Advanced expressive morpho-syntax
9. Rondal & Comblain (in progress)	Claudine (Down syndrome)	27	57	Preoperatory	5 years 4 months		- Correct articulation - Moderately retarded expressive lexical development - Advanced expressive morphosyntax - Moderate difficulties at the discourse level.

Notes.
1.Chronological age in years at beginning of the study.
2. Intellectual quotient according to standard intellectual scales.
3. According to Piagetian criteria.
4. Mental age in years.
5. Mean length of utterance (computed in number of words plus inflectional morphemes).

explanatory possibilities will be examined. They are listed in Table 4.

Table 4. - Explanatory factors for the MR/DS exceptional language cases.

1. Particular educational factors: no
2. Left-hemispheric cerebral dominance: probably necessary.
3. Cognitive functioning (general): no
4. Working memory (phonological loop): possible contributing role.
5. Brain architectural characteristics: likely

2.1 Particular educational factors (unusual language training procedures by parents, educators, teachers, etc.).
Could such influences have determined the state of affairs exemplified in the MR/DS exceptional language cases? The answer is 'no'. I am not implying that a good quality language input is not of utility in language acquisition (see Rondal, 1985, for a favourable outcome). However, there is no indication in the cases reviewed that a particular remedial procedure was responsible for the advanced abilities. Additionally, educational intervention is not known to be particularly effective in compensating for the difficulties of typical MR/DS children with the phonological and morphosyntactic dimensions of language. Moreover, parent-child verbal interactions with MR/DS children have been proven to be basically normal (Rondal, 1978), when by 'normal' it is meant the type of linguistic input and feedback received by NR children at corresponding language levels. Maternal speech addressed to MR/DS language-learning children is adapted to the children's language levels in the same ways (i.e., prosody, articulation, lexicon, semantic structures and contents, morphosyntax, and pragmatics) and to the same extent as mothers' speech to NR children with similar linguistic levels. If adaptations of that sort were the key factor in determining exceptional language abilities in MR/DS subjects, one should observe many more such cases.

2.2 Cerebral hemispheric specialisation for language functions.
Dichotic-listening studies have reported a left ear/right hemisphere advantage for speech sound reception in DS individuals (not found in control groups of NR subjects and MR subjects of other etiologies). DS subjects, however, exhibit the expected right ear/left-hemisphere superiority in speech production. Elliott, Weeks, and Elliott (1987) have suggested that the language problems of DS persons may be related to a dissociation between the cerebral areas responsible for speech perception and production causing difficulties of communication between organic systems that normally overlap, and leaving speech reception in control of the right hemisphere that may not be best equipped to handle this function.

The MR language exceptional subjects for whom relevant data are available (i.e., Françoise, and Curtiss and Yamada's Laura - see Table 1, case no. 2, are both left-hemisphere dominant for language functions (receptive as well as expressive). I have reported (Rondal, 1995) corresponding data for 24 DS adults with typical language abilities for DS (15 males and 9 females, aged 21 to 36 years) in a dichotic-listening study (10 males were right-handed; 5 left-handed; the females were all right-handed) and 19 of the same DS adult subjects (all right-handed) in a dual-task study. A large number of these subjects demonstrated interference between verbalisation and right-hand movements compatible with the hypothesis of a left-hemisphere dominance for speech production. In the dichotic-listening task, three females exhibited a right-ear advantage - from 30 to 70% - (suggesting left-hemisphere dominance). Six males exhibited a right-ear advantage (from 10 to 63%). Retaining those individuals for whom the right-ear advantage was equal to or in excess of 50%, one had two female and one male individuals. These three subjects all demonstrated a positive relative amount of interference in the dual task (suggesting left-hemisphere dominance for speech production). They could be considered homogeneous as to cerebral hemispheric dominance for the speech functions. This is also the case for Françoise (Table 2). However, the language abilities of the above three DS adults were only average for DS persons. Left-hemisphere dominance may be a necessary condition for advanced language development, (outside of early focal brain lesions determining a transfer of the language control to the right hemisphere at little or no functional cost (Eisele, 1991); a situation that theoretically does not concern MR/DS individuals as they are not supposed to present focal brain lesions as a result of their condition). Obviously, however, left-hemisphere dominance cannot be a sufficient condition for exceptional language development in MR/DS people.

2.3 Non-linguistic cognitive factors?
Most MR language-exceptional individuals studied have mental ages (MA) around 5 years. It could be argued that they are 'simply' demonstrating language abilities corresponding to their cognitive level (Moerk, 1994; Bates, 1997, in press). Elizabeth Bates maintains that basic grammatical development is complete by 4-5 years of age (or even before) in NR children. She states (Bates, in

Table 2. - The Françoise (F.)* case (summary of data)
*** French speaking**

1. **CA:** F. was 32 years old at the beginning of the study.
2. **Delay in language development:** F. was producing only one word at 4 years-CA.
3. **Etiology:** Down syndrome (standard trisomy 21: genotype 47,XX, + free 21 in each of the metaphases studied).
4.

IQ (WAIS)	Nonverbal	Verbal
Beginning of study	60	71
End of study	64	70

5. **MA:** (Epreuves Differentielles d'Efficience Intellectuelle - EDEI):
   - Nonverbal: 5 years and 8 months;
   - Verbal: 9 years and 10 months.
6. **Visual perception** (Test of the "Figures Enchevêtrées" de Poppelreuter): normal.
7. **Left-right discrimination** (Test of Head): performance within normal limits.
8. **Visuospatial and computational abilities:** markedly reduced (e.g., standard scores at the WAIS: cubes: 4; object assembly: 1; image completion: 4).
9. **Visuographic abilities** (Complex Figure of Rey -copying from model-; copying cube and houses in perspectives; Bender-Gestalt Test): difficulties with the macrostructure of the drawings; too much attention to irrelevant details; proceeds by copying and juxtaposing small parts of the model; unable to draw according to perspective; on the Bender-Gestalt test, F. scored at the median note for 6-year-old children.
10. **Expressive gesturing** (Immediate imitation of finger and hand-sequential gestures, after Berges and Lezine's Test): F. scored within the 12 year-old range for most gestural sequences.
11. **Attention - concentration** (Barrage subtest of the KLT Test): F.'s scored 22 (out of a possible 90 points); this places her at the lower percentile of the NR adult population.
12. **Operational level** (Piagetian): intermediate between late preoperatory and early operatory.
13. **Episodic memory** (Paired-associate words; Test of the 15 words of Rey; Cued Recall and Selective Reminding Tasks of Buschke; Complex Figures of Rey: drawing from memory (3 minutes after exposure) reduced in comparison with NR adults but satisfactory delayed recall of verbal material (up to 30 minutes) indicating correct trace consolidation; correct but impoverished delayed drawing.
14. **Semantic memory** (Free association; Fluency tasks): F.'s associations are largely idiosyncratic and prevalently of the syntagmatic type; no evidence of prototypical organisation of common semantic categories (e.g., animal, clothes, transportation means, fruits, vegetables, etc.).
15. **Working memory**
    A. Auditory-verbal short-term memory span: 4 units (digits, words, and nonwords)
    B. Visuo-spatial short-term memory span (Block-tapping Test): 4 units (surpassed by 92 % of NR adults; compatible with NR children's level around 5 years).
    C. Visuo-spatial recognition (Delayed Recognition Span Test): span: 5.20 (average span of NR adults: 12.08).
    D. Visual reproduction (Weschler's Clinical Scale): score 4 (very low portion of the NR adult distribution;
NR population mean: 11.42, SD: 2.76).
    E. Basic functioning of phonological loop: demonstrated phonological similarity, word-length, and articulatory suppression effects; spontaneous and active rehearsal (whispering and/or mezza voce).
16. **Sentence span:** 14 words; at times, F. can repeat correctly sentences containing up to 20 words.
17. **L-Max** (Free speech): 50 words.
18. **MLU** (in number of words + inflectional morphemes): 12.24 (SD: 9.65).
19. **Sound perception and discrimination** (ORL, examination): normal (no auditory loss).
20. **Articulatory abilty** (Free speech; Logatomes of Borel-Maisonny): normal.

Table 2. - (continued)

21. **Speech rate** (Free speech measurement): F.'s overt speech rate varies around 200 words per minute (i.e., approximately 3.3 words -12 to 15 phonemes- per second); this is also the speech rate for normal adults; regular DS adults subjects have overt speech rate varying from 37 to 71 words per minutes (i.e., 1 word - 4 to 5 phonemes- per second and less).
22. **Suprasegmental phonology** (Free speech): normal.
23. **Lexical ability -production, referential, comprehension, definition-** (Test de Vocabulaire Actif et Passif; Test des Relations Topologiques; Batterie de l'Aphasie de Liège; Lexical subtest of the Epreuves Différentielles d'Efficience Intellectuelle; Boehm's test of Basic Concepts; Vocabulary subtest of WAIS): productive and receptive levels compatible with nonverbal MA; on the WAIS lexical definition task, the score obtained is one standard deviation below the NR adult population mean.
24. **Expressive morphosyntax** (Free speech analyzed with reference to Halliday's Functional Grammar adapted for French): virtually normal, witness the grammatically correct production of various grammatical types of sentences, including the most structurally complex ones, reflexives constructions, and the correct use of the various obligatory inflectional morphemes.
25. **Receptive morphosyntax** (Specific psycholinguistic tasks): virtually normal; e.g., correct comprehension of subject and object relatives, causal subordinates (with the subordinate clause either preceding or following the main clause), temporal subordinates (with the verbal order of events corresponding to the order of the events in reality or not), declarative affirmative active and passive sentences ranging in plausibility and plausible reversibility, correct use of coreferential mechanism in the case of the anaphoric personal pronouns.
26. **Pragmatic organization** (Free speech): virtually normal conversational skills (e.g., turn-taking, topic distribution, topic continuity, conversational feedbacks and repairs); correctly formulated illocutionary speech acts; correct use of polite forms and indirect requests for action, information, and confirmation.
27. **Discursive organization** (Free speech): discursive organisation (either narrative or descriptive) globally correct; occasional problems with textual cohesion; conjonctive forms such as and, then, but, although, thus, etc., tending to be used more as loose connectors than genuine markers of logical and/or informational dependencies between utterances, phrases, or sentences.
28. **Written language expression** (Free written texts; Dictation): limited and deviant in several respects (e.g., ponctuation marking, conventional orthography, narrative macrostructures, morphological inflections).
29. **Reading and comprehension of written language** (Logatomes and conventional words of Borel-Maisonny; School texts; Written Language Comprehension Task from the Epreuve pour l'Examen du Langage of Chevrie-Müller): reading ability is well established although F. is very slow (which contrasts with her fully speed-appropriate oral verbal ability); written language comprehension is at third-grade level, but demontrates lexical and conceptual limitations.
30. **Metalinguistic abilities**

    A. Phonological awareness (10 subtests orally presented: selecting or producing rhymes, isolating initial or final phonemes in target words, fusing phonemes into words, spelling words, etc.): F. is able to segment common French words into syllabes (but very slowly and at time with some degree of overlapping between neighboring syllabes). She cannot regularly identify separate phonemes in words.

    B. Sentence judgement and repair (for grammaticality and semantic acceptability): F. is able to detect and correct word order errors appearing in grammatically incorrect but semantically appropriate sentences. She is also able to detect and mend grammatically correct but semantically abnormal sentences. However, she did not detect inflectional morphological mistakes.

    C. Grammatical analysis (Active declarative affirmative sentences presented in written form): F. can often identify main verbs (actional as well as nonactional) and grammatical subjects, direct objects, indirect objects, time circumstancial elements, and locative circumstancial elements (in asking the school-type questions: qui - who, quoi - what, à qui - to whom, à quoi - to what, quand - when or où - where). She could perform the above analysis on monopropositional sentences only and, in some cases, on main clauses pertaining to complex sentences but leaving subordinate clauses unanalysed.

Table 2. - (continued)

31. Cerebral hemispheric specialization

A. Dichotic listening (Directed attention procedure): REA (right-ear advantage) or LEA (left-ear advantage) calculated from the following formula computed for each ear:

$$\text{Dichotic-listening score} = \frac{(30\text{-}E1)}{30} * 100$$

Where 30 is the number of syllabes presented to each ear and E1 the number of intrusion errors.
F.'s REA = 63%, suggesting LHD (left-hemisphere dominance) for speech reception.

B. Dual-task study (Finger-tapping task combined with sound-shadowing): Relative amount of interference averaged per second (RAI index) evaluated with a formula comparing experimental steps.
F.'s RAI = +4.05, indicative of an interference between verbalization and finger-tapping that is more marked for the right hand; suggesting LHD for speech production.

Table 3. - Claudine - Study in progress
*** French speaking**

1. **CA:** C. was 27 years old at the beginning of the study.
2. **Etiology:** Down syndrome (standard trisomy 21).
3. **IQ** (WAIS): nonverbal 57, verbal 61.
4. **MA** (EDI) nonverbal: 5 years and 4 months; verbal: six years and 3 months.
5. **Nonlanguage cognition:** important weaknesses in spacial, numerical and time cognition; preoperatory level on Piagetian mesures
6. **Working memory:** A-V/ST span: 4; V-S/STM span:4.
7. **Speech rate** (articulation speed): below NR but >to typical DS individuals
8. **Oral language production:**
   Lexicon: EVP 68%; not very different from typical DS adult individuals;
   - MLU (words + inflection morphemes); 15.39;
   - Grammatically correct expression of temporal, casual, and relative subordinates in free speech; her speech on the whole is more parataxic than that of Françoise.
9. **Written language:**
   - Correct writing and reading abilities, albeit very slowly;
   - Correct marking of subject - main verb or subject - auxiliary concord; correct use of the obligatory inflections on the nouns, verbs, adjectives, and pronouns.
10. **Metalinguistic ability:** seemingly clear phonological awareness.

press): 'The amount and type of cognition required to learn a grammar cannot be more than the amount and type of cognition that is available to healthy normal children between 1.5 and 3 years of age, for that is the period in which the bulk of grammatical development takes place' (p. 3). From such a standpoint, one should expect MR/DS individuals with MAs of 4 or 5 years to exhibit well-developed formal language abilities.

If general cognition at 4-5 years-MA were a sufficient condition for explaining advanced formal language abilities, typical MR/DS subjects with such MAs (and there are many) should similarly exhibit well-developed morphosyntactic skills. Unfortunately for them and for the above claim, typical MR/DS individuals as a rule remain much below complete grammatical development during their whole lives, despite often receiving systematic and longstanding language intervention.

Alternatively, if one does not accept the idea that grammatical development is complete by 4-5 years of age but insists instead that it goes on until 9-10 years for some complex syntactic structures, then the levels reached by the exceptional MR/DS individuals become impossible to explain in relying solely on general cognitive variables. Indeed, these individuals exhibit grammatical levels much beyond what would be considered normal development around 4-5 years MA in this alternative hypothesis. The general cognition hypothesis is contradicted by the data either from the typical MR/DS subjects or from the language-exceptional MR/DS individuals, depending on how one wants to set the timing for completion of grammatical development in NR children. In my opinion, the language-exceptional MR/DS individuals have more to rely on than a cognitive level of 4-5 years MA. They have at their disposal a language specific ability (grammatical) that has been spared in spite of their pathology. This ability

is also available to young NR children (around 20-24 months) when they start developing grammar. It is largely lacking in typical MR/DS individuals.

Bates (in press) states 'In order to prove that cognitive abilities are unnecessary for language, - [my note: it would be better to speak of grammar or even more narrowly of morphosyntax rather than of language, for there are reasons to believe that cognition is necessary for the conceptual aspects of language in order to develop and function; and I do not believe that this point is controversial] - we would have to find a case in which grammar is acquired in the absence of the specific cognitive abilities that two-year olds have at their disposal during the language learning process (p.3)'. It is true that such a demonstration has not been provided. The reverse situation is currently observed, however. All typical moderately and severely MR individuals reach and go beyond 2 years MA, but, as said, they fall short of developing full grammar. Early cognitive development may supply the necessary basis for grammatical development, but it cannot be a sufficient condition. Needed too is a specific device responsible for the grammatical operations.

The grammatical ability referred to does not have to be innate in the representational sense out of any logical or biological necessity. As will be discussed later in the paper, it does imply, however, the existence of innate architectural constraints (to employ Elman et al.'s, terminology, 1996), i.e., the innate organisation of the processing system that must deal with linguistic, and particularly grammatical, representations. Karmiloff-Smith et al. (1995) suggest that there are domain specific predispositions for analysing language stimuli which with language experience, become increasingly specialised and interconnected. As normal development proceeds, a process of 'modularization' (p. 23) gives rise to a modular-like organisation (weaker than the Fodorian view of modules - Fodor, 1983). These types of modules can be said to be more made than born (Bates et al., 1988). But, judging from the vantage point of the MR literature, fully efficient language modularization, to continue with Karmiloff-Smith's terminology, does not occur in typical MR/DS subjects despite some cognitive, lexical, and pragmatic acquisitions. This again suggests that something else is needed to bring about the modularization process that may be characteristic of advanced language development and functioning.

As suggested, early cognitive functioning is relevant for early morphosyntactic development. Supportive evidence can be found in several exceptional MR/DS cases. Françoise as well as Christopher, O'Connor and Hermelin's subject, Laura, Curtis and Yamada's subject, and FF, Vallar and Papagno's subject - i.e., those language exceptional individuals for whom we have developmental histories - were markedly delayed in language, onset. Françoise was producing only one word (/to/ for couteau, i.e., in French) at 4 years CA, even worse than many typical DS children. She developed her formal language abilities between approximately 5 and 10 years. WS children are severely delayed in early language development. It is only as adolescents that they exhibit the advanced formal abilities documented (Singer et al., in press). Bates (in press) suggests that it is only when they have a vocabulary size and general cognitive level comparable to those of NR two-years olds that the grammar of the WS individuals 'gets off the ground.' The above observations suggest that a cognitive-semantic basis amounting to what is known by the moderately and severely MR child around 5 years-CA and to the NR child around 20-24 months is needed for the grammatical component to start working when such a component is indeed available.

2.4. What about working memory (Baddeley, 1990) in relation with exceptional language development in MR/DS individuals?

Françoise, Claudine, as well as Vallar and Papagno's FF, have an auditory-verbal short-term memory (AV-STM) span of 4 or more (FF, in particular). The AV-STM spans of typical DS persons do not exceed 3 or 4 units. They are impaired in the functioning in the phonological loop of their working memory system (Jarrold & Baddeley, 1997). They also exhibit very limited to non-existent private speech and rehearsal abilities (Comblain, 1996). Françoise, Claudine, and FF's AV-STM spans certainly were lower at the time of their language development than recent estimates. As said, Françoise developed her particular language abilities between 5 and 10 years CA. In NR children, immediate verbal memory span increases from a mode of 4 items around 5 years CA to 5 items around 7 years, and 5.5 items around 10 years (Mackenzie & Hulme, 1987). In MR/DS children, Mackenzie and Hulme (1987) observed little increase in digit span over a 5-year period in their DS group (from about 3 items at 11 years CA to 3.5 items at 16 years) as well as in a group of moderately and severely MR children of other etiologies (from about 3.5 items at 11 years CA to 4 items at 16 years). Vallar and Papagno (1993) proposed that FF's better AV-STM (i.e., 5.75 items) explains her better formal language abilities. Such a strong suggestion is not convincing.

However, a positive contribution of AV-STM, due to a better functioning of the phonological loop of the AV-STM system, cannot be ruled out in MR/DS language-exceptional subjects. Françoise, Claudine, as well as FF, exhibit normal-like processes of working memory when recalling verbal material (e.g., word-length, phonological similarity, and Brown-Peterson effects). They rely on rehearsal strategies based on semi-private speech. Their speech rate is normal or close to normal in contrast to that of typical DS subjects (Rondal, 1995). This is an important indication as Baddeley (1990) specifies the content of the phonological loop of working memory as 'the number of items of whatever length that can be uttered in about two seconds' (p.74).

Correlative data published by Gathercole and associates (e.g.,Gathercole, 1995; Gathercole & Baddeley, 1993; Gathercole, Willis, Emslie, & Baddeley, 1992) suggest that phonological short-term memory may contribute to word learning during normal development at least until approximately 4 years. By that age, other factors such as the way in which lexical items are integrated into the existing lexicon and the way the lexicon is organised, may exert more of an effect than in earlier stages of word learning. But no model has been proposed specifying exactly how, when, and to what extent, the phonological loop mediates early lexical or morphosyntactic development.

Grant et al. (in press) attribute the advanced levels of expressive and receptive vocabulary documented in a group of 17 individuals with WS (aged 8 to 35 years) to their relatively preserved phonological short-term memory. For these subjects, however, as well as for the language-exceptional MR/DS mentioned above, and even for the NR subjects of Gathercole and associates, the direction of causality could be the reverse, i.e., better lexical abilities gradually permitting enhanced short-term memory functioning; an alternative hypothesis that has not received sufficient attention in the working memory literature.

Let us consider the Françoise case again to pursue the discussion and set neat limitations on the possible contribution of AV-STM to exceptional formal language development in MR/DS subjects. As said, Françoise's STM span is 4. Her sentence span is 14 words. At times, she can repeat correctly sentences containing up to 20 words. This is normal functioning according to data reported by Butterworth, Campbell, and Howard (1986). Françoise made few word order errors on sentences containing more than 14 words. Most of her errors were omissions and trivial word substitutions. So were also the typical errors of Butterworth et al 's university students when requested to recall sentences 15-20 words long. Françoise's immediate recall performance is in sharp contrast with that of typical DS subjects (Rondal, 1995). The latter individuals cannot repeat correctly sentences containing more than 7 or 8 words at best. They frequently omit major sentence constituents. Additionally, Françoise had no difficulty in correctly interpreting (center-) embedded subject and object relatives when the relative pronouns and their co-referring nouns were separated by several words. Neither did she experience particular problems when requested to establish pronominal co-reference across sentences in paragraphs with pronouns and co-referring nouns separated by up to eight words. It seems reasonable to conclude that the contribution of Françoise's immediate phonological memory to sentence production and comprehension is sharply limited.

2.5. Variation at brain level.

My guess is that the major determinant of the impressive morphosyntactic and phonological differences observed between typical and exceptional MR/DS subjects operates at brain level. The macroscopic brain structures devoted to the formal aspects of language (as opposed to the more semantic ones) are probably spared to a large extent in those MR/DS individuals with exceptional language abilities. They are damaged and only poorly operational in regular MR/DS subjects. I will suggest a likely reason for this state of affairs later in the paper. But let me first consider and reject an alternative explanatory hypothesis. It can be formulated as follows. Suppose one assumes something like representational nativism to be true, he could argue that what is spared in the language-exceptional MR/DS cases is not so much the devoted brain structures than (unconscious) linguistic notions ascribable to innate properties of the human mind. One would then be led to suggest that the basic language differences between exceptional and regular MR/DS subjects lie at the level of the micro-circuitry of the brain. This is at that level that nativists (e.g., Pinker, 1994) tend to locate innate representational language knowledge.

Such an hypothesis must be rejected for the following reasons. First, representational nativism can be challenged on logical and neurobiological grounds (see Elman et al., 1997, for a detailed exposition). It is becoming clearer that linguistic representations are constructed on the basis of children's experiences with language. Bates et al. (1996) agree that synaptic connections are the likely depository structures for linguistic representations because that is the only brain level

with the coding power for higher-order cognitive outcomes. The difference between this view and representational nativism is that the devoted synaptic connections here are viewed as modifiable and actually modified by linguistic experience. Second, there is no reason to believe that language exceptional and typical MR/DS subjects differ at the level of synaptic connectivity as a result of genetic differences. And, third, if it were the case exceptional language cases could be found only in genetic syndromes (which does not happen - see Table 1).

As suggested, language-exceptional MR/DS individuals enjoy preserved brain macro-structures devoted to the treatment of formal aspects of language. This suggestion may be related to Elman et al.'s (1996) concept of architectural constraints at the brain level. This notion refers to the innate structuring of the brain information-processing system devoted to the acquisition and use of linguistic representations. As Bates et al. (1996) state 'In neural networks, some forms of knowledge can only be realised or acquired in a system with the right structure (the right number of units, number of layers, types of connectivity between layers, etc.)'(p. 3). Specifying further the architectural constraints in real brains, Elman et al. (1996) distinguish three sub-levels; the basic computing units (neurons, neurotransmitters, etc.); the local architectures (number and thickness of neuronal layers, density of different cell types within layers, type of neuronal circuitry, etc.); and the global architecture (including the afferent and efferent neural pathways).

Correctly organised brain macro-structures owe much to the interplay of what Elman et al. (1996) label 'chronotopic constraints'. This includes constraints on the number of cell divisions taking place in neurogenesis, relative differences in timing between brain subsystems, differences in synaptic growth according to brain areas and functions, etc. My suggestion is that language-exceptional and typical MR/DS subjects markedly differ as to the architectural and chronotopic characteristics of brain development. Results of pathological studies of the brain of DS persons reveal major anomalies, among which arrested maturation of neurons and synapses some time around birth, reduced brain weight, reduced neuronal densities, decreased synaptic density and presynaptic length, hypoplasia of the frontal lobes, narrowed superior temporal gyri, delayed myelination of nerve fibers affecting long association and intercortical fibers between frontal and temporal lobes, hypothalamic and hippocampal abnormalities, and diminished size of brain stem and cerebellum have been reported (Wisnieswky, Kida, & Brown, 1996; Nadel, 1996).

To date there have been few functional studies of brain function in DS individuals. Devinsky et al. (1990) reported normal EEG activity in young DS adults, but McAlaster (1992) reported abnormal development of EEG profiles in DS subjects. Horwitz et al. (1990)'s PET (positron emission tomography) scan study of cerebral metabolic patterns in young DS adult subjects reveal smaller correlations for region-pairs within and between frontal and parietal lobes. One brain region particularly affected is the inferior frontal gyrus including Broca's area. The thalamus shows smaller correlations with the temporal regions in the DS group compared to controls. Shapiro, Haxby, and Grady (1992), also using PET scan, report a corresponding disruption of neuronal interactions between frontal and parietal lobes, possibly including Broca's area.

Bates et al. (1996) suggest that left-hemisphere specialisation for language in humans depend on the architectural constraints specified above. They further claim that studies of infants with focal brain injury demonstrate that the temporal (but not the frontal) region of the left hemisphere is specialised at birth. Later in life, the temporal and the frontal regions of the left hemisphere play a major role in language production and comprehension in the great majority of NR persons (a small percentage of NR adults exhibit a right-hemisphere dominance or a mixed specialisation for the language functions).

The particular brain areas responsible for the expressive and receptive treatment of the formal language aspects in normal adults are becoming better known. According to Damasio and Damasio (1989, 1992), they involve the posterior perisylvian sector of the left-cerebral hemisphere, including the basal ganglia, with respect to the processing of speech sounds, phoneme assembly into words, and selection of word forms; the anterior perisylvian sector of the left hemisphere, including the basal ganglia, with respect to receptive and expressive morphosyntax. Stowe et al.'s (1994) review of PET scan studies points to the involvement of the left-cerebral hemisphere extrastriate cortex and superior temporal cortex in lexical access; Brodmann's areas 41, 42, and mid-Brodmann's 22, in phonological processing; the superior temporal cortex in syntactic processing; and the inferior frontal gyrus, the mid and inferior temporal gyri, Brodmann's area 8, and the temporal poles, in discourse comprehension.

It is reasonable to assume that the brain problems typical of DS individuals seriously undermine the development and functioning of the language brain structures. Particularly detrimental is probably the

slowing down of synaptic growth around birth (Nadel, 1986). The consequence is that typical DS subjects do not develop the necessary brain architecture for accommodating language stimuli in such a way as to build grammatical knowledge. It is likely also that the plasticity which characterises normal brain development allowing functional reorganisation in case of early focal brain injury - is not open to DS infants for reasons of their genetic pathology and the sharp restrictions that it imposes on brain genesis and epigenesis.

The language-exceptional DS subjects escape the above fate for reasons that may be related to the phenotypic effects of genetic variation in this syndrome. The same suggestion is valid for other genetic syndromes as well. Geneticists agree that there is substantial variation at the genetic level between people within genotypic categories such DS, WS, Fragile-X syndrome (FXS), and other genetic causes of mental retardation (Dykens, 1995). Most genetic influences on phenotypes are not discrete and the inheritance patterns may be a blend between single gene and polygenic influences (Smith, Pennington, & DeFries, 1996). Complex traits show a quantitative variation in their presentation. The major sources of variation number four. There may be, first, a single major gene involved in a trait and it may show variable penetrance. Penetrance is defined by the proportion of individuals with the susceptible gene(s) having a given disorder. Second, there may be variable expression of a single major gene or of a number of genes involved, due to the modifying influences of other genes or environmental factors, leading to differences in phenotypes., a major gene may have many possible mutations (alleles) that may also have different phenotypes. And, fourth, imprinting effects, i.e., variability of gene expression associated with the parental origin of the excess genetic material.

Genetic research is yielding more precise gene identification and phenotypic mapping of chromosome 21, the chromosome involved in Trisomy 21 (T21). Korenberg et al. (1994) suggest that DS is a contiguous gene syndrome. This augurs against a single DS chromosomal region responsible for most of the DS phenotypic features. DS and its phenotypes are accurately thought of as the result of the over-expression and subsequent interactions of a subset of the estimated 1,000 to 1,700 genes located on chromosome 21. Korenberg et al. (1994) have constructed a phenotypic map including 25 features considered typical of DS. They assign a region of 2-20 mega-bases between regions p11.2 and 22.3 on the distal part of the long arm of chromosome 21, as likely to contain the genes responsible for the DS phenotypes. This conception of the genotype-phenotype relationship in DS is consistent with central characteristics of T21, such as the rich variety of phenotypes and the variability in both penetrance and expression of the phenotypic features. It is conceivable that an important inter-individual variation exists at brain level in the language areas (as well as in other areas) of DS persons, consequent on genetic variations. One may imagine a sort of Gaussian curve of formal language abilities in DS people, or better several Gaussian curves, one for each formal dimension. The exceptional cases documented (and others to come may be) would occupy the extreme right portion of such distributions of formal language abilities.

3. The brain-genes perspective defined above has the advantage of proposing one single explanation for the range of variations observed in the language of typical MR/DS people and the extremes of such variations in the language-exceptional cases. It may also be applied to behavioural and brain differences across syndromes such as Down syndrome, Williams syndrome, and Fragile-X syndrome; a large number of other genetic syndrome awaiting systematic phenotypical studies (Dykens, 1995). Rondal and Edwards (1997) have reviewed a number of research works on the language abilities of WS and FXS individuals. When the results of these studies are compared with each other and with what is known of the language of typical DS subjects, a feature distribution like the one illustrated in Table 5 is revealed.

WS is a multisystem developmental disorder caused by the deletion of contiguous genes at locus 7q11.23. Hemizygosity of the elastin (ELN) gene accounts for the vascular and connective tissue abnormalities observed in WS patients. However, the genes that contribute to other features of the syndrome, such as infantile hypercalcemia, dysmorphic facies, and cognitive defects (between mild and moderate mental retardation) remain to be identified. FXS is an X-linked disorder passed on through generations. It is caused by a null mutation at the FMR-1 gene in which the levels of protein in mRNA (messenger ribonucleic acid) are greatly reduced. At the DNA (desoxyribonucleic acid) level, it is characterised by abnormal repetitions of a trinucleotide sequence (either cytosine-cytosine-guanine or cytosine-guanine-guanine). Most males with FXS are moderately to severely mentally retarded. One fifth (considered to be nonpenetrant) are of average intelligence. Approximately, one third of the FXS females are affected with a phenotypic variant of the syndrome. They exhibit learning difficulties. Some are impaired with mild to moderate MR. The rest of the carrier females are unaffected. Yet they may transmit the disorder to their children. Surveys of

Rondal Table 5. - Three MR language syndromic profiles

	Syndromes		
	Down	Williams	Fragile-X (affected males)
Language aspect			
Phonetico-phonological	— —	+	— —
Lexical	—	++	+
Thematic semantic	+	+	?
Morphosyntactic	— —		—
Pragmatic	+	— —	—
Discursive	— —	+	—

Key. +(+): relative strength; —(—): relative weakness; ?: insufficient data available.

MR populations (Webb et al., 1986) suggest that FXS accounts for 2 to 7 percent of MR amonmales. FXS ' prevalence in the general population is between 0.73 and 0.72 per 1000 males.

Pioneering work by Bellugi and associates at the Salk Institute for Biological Studies suggest that functional differences between WS and DS individuals correspond to (and are probably caused by) syndromic variation at brain level. Bellugi et al. (1990) compared the neurological profiles of WS and DS adolescents matched for CA and IQ. The WS subjects demonstrated generalized hypotonia, tremor, midline balance problems and oral-motor and motoric abnormalities, suggestive of cerebellar dysfunction. DS adolescents showed minimal hypotonia, little evidence of cerebellar signs, and better performance on oromotor functions. Both groups exhibited equal degrees of microcephaly, cerebral hypoplasia, reduced cerebral volume, and decreased myelination; but overall brain shapes of each group proved distinct. DS brains exhibit important degrees of hypofrontality whereas WS individuals have decreased posterior width with reduction in size of the forebrain posterior to the rolandic sulcus, i.e., the parietal, posterior temporal, and occipital cortical regions, and narrowing of the corpus callosum anterior to the splenium. WS individuals show elongated posterior to anterior length compared to normal brains. Hypofrontality of neocortex in DS subjects together with reduction in frontal lobe projections from the corpus callosum was further demonstrated in a magnetic resonance imagery study by Wanet al. (1992). These authors relate this neuroanatomical indication to a profile of frontal lobe dysfunction in DS corresponding to poor verbal fluency, perseverative tendencies, and greater difficulty on tasks requiring flexible problem-solving strategies. DS subjects, however, seem to have relatively preserved basal ganglia and diencephalic structures. In contrast, WS subjects have better frontal and temporal limbic structures (Jernigan et al, (1993).There is also evidence in WS of a dysregulation of the control of neuronal and glial numbers, as illustrated by increased cell packing density at the cytoarchitectonic level (Galaburda et al., 1994). This may reflect an interference with naturally occurring cell death and the presence of neurotrophic factors (possibly linked to abnormal extracellular calcium levels).

The cerebellar volume in DS subjects is approximately 77 per cent of the equivalent volume in young normal controls, versus 99 per cent in WS subjects. Although cerebellar size is intact and neocerebellum largely preserved in WS (Wang, 1992), some other neurological findings are suggestive of cerebellar dysfunction. The posterior fossa structures of the WS and DS subjects were further examined by Bellugi et al. (1990), leading to the identification in WS subject of an anomalous pattern, with neocerebellar vermal lobules showing hyperplasia in the context of low-normal paleocerebellar vermal development and significantly reduced forebrain size. Such an aberrant cerebrum/cerebellum volume ratio could serve to distinguish WS neurologically from other syndromes such as DS or autism (Courchesne et al. 1988). Bellugi et al. (1990) speculate (following suggestions by Leiner, Leiner and Dow, 1986 on the possible role of human neocerebellar structures in mental and linguistic functions) that the

observed hyperplasia of specific vermal lobules in the context of cerebral maldevelopment may be related to the language profile of their WS subjects. These subjects demonstrate relatively preserved syntax and advanced referential lexical abilities, whereas the DS subjects' language profiles are flatter. Bellugi et al. (1990) further remark that their WS subjects are behaviourally grossly similar to unilateral right-hemisphere damaged (normal) adults whereas the DS individuals are more like left-hemisphere damaged aphasics, demonstrating language impairment and a marked tendency to a global processing of the information. This is intriguing from a neuropsychological point of view given that in WS as well as in DS no focal brain damage is considered to exist. The curtailment of the dorsal parietal and posterior temporal areas of the brain in WS subjects, together with the thinning of portions of the corpus callosum, may be directly relevant to their visuo-spatial deficits (Galaburda et al., 1994), and indirectly perhaps to the dissociation between auditory-verbal and visuo-spatial short-term memory systems. WS subjects indeed have better preserved AV- than VS-STM, whereas the converse is true for many DS subjects (Jarrold & Baddeley, 1997). Similarly, the relatively preserved size of the frontal and most of the temporal lobes in WS is consistent with the relative preservation of formal linguistic capacity in many of these subjects.

According to Galaburda et al. (1994), several features of cortical architectonic differentiation in WS (e.g., increased cell packing density, horizontal disposition of neurons and other anomalous layering of the cortex, excessive number of subcortical neurons, immature vascular development, weak myelination of fibers) suggest an arrest in neuronal development between the end of the second trimester and the second year of life. For Down syndrome, the estimated time of marked slowing down in neuronal development is around birth. It is tempting to speculate that the 6-month to 1-year difference in neural epigenesis between DS and WS is in relation with the better preservation of the formal language organisation in the latter individuals. This may constitute an indication that brain epigenetic development during the first six months or the first year of life significantly affects the development of dedicated architectural structures. Pursuing careful investigations in the separation of higher cortical functions in well-defined genetically based syndromes will shed interesting lights on the neural systems subserving the language functions.

4. The preceding analysis encourages the belief that considerable value is to be gained by the creation of a shared platform for a heuristic and theory-motivated dialogue across such disciplines as genetics, linguistics, language acquisition, language disorders, brain imaging and the cognitive sciences in general, with regard to the genetic dysphasies, among which DS represents an important and historically primordial category. The research logic is clear. It consists in (1) specifying the behavioral phenotypes regarding various aspects of language; (2) determining the specific neuroanatomy and brain functions underlying these phenotypes; and (3) identifying the specific genotypes responsible for the individual differences and influencing the language functions involved. A major research avenue opens leading towards a neurobiological psycholinguistics. Progress along this path will lead to a better understanding of the intrinsic nature of the language profiles in MR/DS persons.

Acknowledgements:
I am grateful to Elizabeth Bates for insightful discussions and exchanges on the theme of this paper as well as for supplying me with several papers still unpublished or currently in press.

Correspondence:
Laboratoire de Psycholinguistique, Boulevard du Rectorat, 5, B-32, 4000, Liege, Belgium.
E-mail: ja.rondal@ulg.ac.be

References

Anderson, E., & Spain, B. (1977). *The child with spina bifida.* London:Methuen.

Baddeley, A. (1990). *Human memory: Theory and practice.* Hove: Erlbaum.

Bates, E. (1997, May). *Personal communication.*

Bates, E. (in press). On language savants and the structure of mind. *International Journal of Bilingualism.*

Bates, E., Bretherton, I., & Snyder, L. (1988). *From first word to grammar. Individual differences and dissociable mechanisms.* Cambridge, UK: Cambridge University Press.

Bates, E., Elman, J., Johnson, M., Karmiloff-Smith, A., Parisi, D., & Plunkett, K. (1996, May). *On innateness* San Diego: University of California, Center for Research in Language, Technical report 9602.

Bellugi, U. (1997, March). Personal communication.

Bellugi, U., Bihrle, A., Jernigan, T., Trauner, D., & Doherty, S. (1990). Neuropsychological, neurological and neuroanatomical profile of Williams syndrome. *American Journal of Medical Genetics Supplement*, 6, 115-125.

Bellugi, U., Marks, S., Bihrle, A., & Sabo, H. (1988). Dissociation between language and cognitive functions in Williams syndrome. In D. Bishop and K. Mogford (Eds.), *Language development in exceptional circumstances* (pp. 177-189). London: Churchill Livingstone.

Butterworth, B., Campbell, R., & Howard, D. (1986). The uses of short-term memory: A case study. *Quarterly Journal of Experimental Psychology*, 38, 705-737.

Comblain, A. (1996). Le fonctionnement de la mémoire à court-terme auditivo-vocale dans le syndrome de Down: Implications pour ler modèle de mémoire de travail. *Approches Neuropsychologiques des Apprentissages de l'Enfant*, 39-40, 137-147.

Courchesne, E., Yeung-Courchesne, R., Press, G., Hesselink, J., & Jernigan, T. (1988). Hypophasia of cerebellar vermal lobules VI and II in autism. *New England Journal of Medicine, 318,* 1349-1354.

Cromer, R. (1991). *Language and thought in normal and handicapped children.* London: Blackwell.

Curtiss, S. (1989). Abnormal language acquisition and the modularity of language. In F. Newmeyer (Ed.), *Linguistics: The Cambridge survey* (vol. 2, pp. 96-116). Cambridge, UK: Cambridge University Press.

Damasio, A., & Damasio, H. (1992). Brain and language. *Scientific American,* September, 89-95.

Damasio, H., & Damasio, A. (1989). *Lesion analysis in neuropsychology.* Oxford, UK: Oxford University Press.

Devinsky, O., Sato, S., Conwit, R., & Schapiro, M. (1990). Relation of EEG alpha background to cognitive function, brain atrophy and cerebral metabolism in Down's syndrome. *Archives of Neurology, 47,* 58-62.

Dykens, E. (1995). Measuring behavioral phenotypes: Provocations from the "new genetics". *American Journal on Mental Retardation, 99,* 522-532.

Eisele, J. (1991). Selective deficits in language comprehension following early left and right hemisphere damage. In I. Pavao Martins, A. Castro-Caldas, H. Van Dongen & J. Van Hout (Eds.), *Acquired aphasia in children* (pp. 225-238). Boston, MA: Kluwer.

Elliott, D., Weeks, D., & Elliott, C. (1987). Cerebral specialization in individuals with Down syndrome. *American Journal on Mental Retardation, 92,* 263-271.

Elman, J., Bates, E., Johnson, M., Karmiloff-Smith, A., Parisi, D., & Plunkett, K. (1996). *Rethinking innateness: A connectionist perspective on development.* Cambridge, MA: MIT Press.

Fodor, J.A. (1983). *The modularity of mind.* Cambridge, MA: MIT Press.

Galaburda, A., Wang, P., Bellugi, U., & Rossen, M. (1994). Cytoarchitectonic anomalies in a genetically based disorder: Williams syndrome. *Cognitive Neuroscience and Neuropsychology, 5,* 753-757.

Gathercole, S. (1995). Is nonword repetition a test of phonological memory or long-term memory knowledge? It all depends on the nonwords. *Memory and Cognition, 23,* 83-94.

Gathercole, S., & Baddeley, A. (1993). *Working memory and language.* Hillsdale, NJ: Erlbaum.

Gathercole, S.E., Willis, C., Emslie, H., & Baddeley, A.D. (1992). Phonological memory and vocabulary development during the early school years: A longitudinal study. *Developmental Psychology, 28,* 887-898.

Grant, J., Karmiloff-Smith, A., Gathercole, S., Paterson, S., Kowlin, P., Davies, M., & Udwin, O. (in press). Phonological short-term memory and its relation to language in Williams syndrome. *Cognitive Neuropsychiatry.*

Hadenius, A., Hagberg, B., Hyttnas-Bensch, K., & Sjogren, I. (1962). The natural prognosis of infantile hydrocephalus. *Acta Pediatrica, 51,* 47-118.

Horwitz, B., Schapiro, M., Grady, C., & Rapoport, S. (1990). Cerebral metabolic pattern in young adult Down's syndrome subjects: Altered intercorrelations between regional rates of glucose utilization. *Journal of Mental Deficiency Research, 34,* 237-252.

Jarrold, C., & Baddeley, A. (1997). Short-term memory for verbal and visuospatial information in Down's syndrome. *Cognitive Neuropsychiatry, 2,* 101-122.

Jernigan, T. Bellugi, U., Sowell, E., Doherty, S., & Hesselink, J. (1993). Cerebral morphologic distinctions between Williams and Down's syndromes. *Archives of Neurology, 50,* 186-191.

Karmiloff-Smith, A., Grant, J., Berthoud, I., Davies, M., Howlin, P., & Udwin, O. (1997). Language and Williams syndrome. How intact is intact? *Child Development, 68,* 274-290.

Karmiloff-Smith, A., Klima, E., Bellugi, U., Grant, J., & Baron-Cohen, S. (1995). Is there a social module? Language, face processing, and theory of mind in individuals with Williams syndrome. *Journal of Cognitive Neuroscience, 7,* 196-208.

Karmiloff-Smith, A., Tyler, L., Voice, K., Sims, K., Udwin, O., Howlin, P., & Davies, M. (unpublished). *Linguistic dissociations in Williams syndrome: Evaluating receptive syntax in on-line and off-line tasks.* London: Medical Research Council, Cognitive Development Unit.

Korenberg J., Chen, X., Schipper, R., Sun, Z., Gonsky, R., Gerwehr, S., Carpenter, N., Daumer, C., Dignan, P., Distche, C., Graham, J., Hugdins, L., McGillivray, B., Miyazaki, K., Ogasawara, N., Park, J., Pegon, R., Pueschel, S., Sack, G., Fay, B., Schuffenhauer, S., Soukup, S., & Yamanaka, T. (1994). Down syndrome phenotypes. The consequences of chromosomal imbalance. *Proceedings of the National Academy of Sciences, 91,* 4997-5001.

Leiner, H., Leiner, A., & Dow, R. (1986). Does the cerebellum contribute to mental skills? *Behavioral Neuroscience, 100,* 443-454.

Mackenzie, S., & Hulme, C. (1987). Memory span development in Down's syndrome, severely subnormal and normal subjects. *Cognitive Neuropschology, 4,* 303-319.

McAlaster, R. (1992). Postnatal cerebral maturation in Down's syndrome children: A developmental EEG coherence study. *International Journal of Neuroscience, 65,* 221-237.

Moerk, E. (1994, October). *Personal communication*

Nadel, L. (1986). Down syndrome in neurobiological perspective. In C. Epstein (Ed.), *The neurobiology of Down syndrome* (pp. 239-251). New York: Raven Press.

Nadel L. (1996). Learning, memory and neural function in Down syndrome. In. J.A. Rondal, J. Perera, L. Nadel & A. Comblain (Eds.), *Down's syndrome: Psychological, psychobiological and socio-educational perspectives* (pp. 21-42). London: Whurr.

O'Connor, N., & Hermelin, B. (1991). A specific linguistic ability. *American Journal on Mental Retardation, 95,* 673-680.

Pinker, S. (1994). *The language instinct.* New York: Morton.

Rondal, J. A. (1978). Maternal speech to normal and Down's syndrome children matched for mean length of utterance. In C. Meyers (Ed.), *Quality of life in the severely and profoundly retarded people: Research foundations for improvement* (pp. 193-265). Washington, D.C.: American Association on Mental Deficiency.

Rondal, J. A. (1985). *Adult-child interaction and the process of language acquisition.* New York: Praeger.

Rondal, J. A. (1994a). Exceptional cases of language development in mental retardation: The relative autonomy of language as a cognitive system. In H. Tager-Flusberg (Ed.), *Constraints on language acquisition: Studies of atypical children* (pp. 155-174). Hillsdale, NJ: Erlbaum.

Rondal, J. A. (1994b). Exceptional language development in mental retardation: natural experiments in language modularity. *Cahiers de Psychologie Cognitive, 13,* 427-467.

Rondal, J. A. (1995). *Exceptional language development in Down syndrome: Implications for the cognition-language relationship.* Cambridge, MA: Cambridge University Press.

Rondal, J. A., & Edwards, S. (1997). *Language in mental retardation.* London: Whurr.

Seagoe, M. (1965). Verbal development in mongolism. *Exceptional Children, 6,* 229-275.

Shapiro, M., Haxby, J., & Grady, C. (1992). Nature of mental retardation and dementia in Down syndrome: Study with PET, CT, and neuropsychology. *Neurobiology of Aging, 13,* 723-734.

Singer, N., Bellugi, U. Bates, E., Jones, W., & Rosen, M. (unpublished). C*ontrasting profiles of language development in children with Williams and Down syndrome.* San Diego: University of California, Department of Psychiatry.

Smith, N., & Tsimpli, I.M. (1995). *The mind of a savant. Language learning and modularity.* Oxford, UK: Blackwell.

Stowe, L., Wijers, A., Willemsen, A., reuland, E., Paans, A., & Vaalburg, W. (1994). PET studies of language: An assessment of the reliability of the technique. *Journal of Psycholinguistic Research, 23,* 499-527.

Tew, B. (1979). The *"cocktail party syndrome"* in children with hydrocephalus and spina bifida. British Journal of Disorders of Communication, 14, 89-101.

Vallar, G., & Papagno, C. (1993). Preserved vocabulary acquisition in Down's syndrome: The role of phonological short-term memory. C*ortex, 29,* 467-483.

Wang, P. (1992, June). *Relationship of neuropsychological and neurobiological profiles in Williams' and Down's syndromes.* Paper presented at the Symposium "Two genetic syndromes of contrasting cognitive profiles" of the American Psychological Society, San Diego, CA.

Wang, P., Doherty, S., Hesselink, J., & Bellugi, U. (1992). Callosal morphology concurs with neuropathological findings in two neurodevelopmental disorders. *Archives of Neurology, 49,* 407-411.

Webb, T., Bundey, S., Thake, A., & Todd, J. (1986). Population incidence and segregation ratios in Martin-Bell syndrome. *American Journal of Medical Genetics, 23,* 573-580.

Wisnieski, K., Kida, E., & Brown, T. (1996). Consequences of genetic abnormalities in Down's syndrome on brain structure and function. In. J.A. Rondal, J. Perera, L. Nadel & A. Comblain (Eds.), *Down's syndrome: Psychological, psychobiological and socio-educational perspectives* (pp. 3-19). London: Whurr.

Yamada, J. (1990). *Laura. A case for the modularity of language.* Cambridge, MA: MIT Press.