

OUTCOME OF SEVERE RECEPTIVE LANGUAGE DISORDER

The clinical and familial characteristics of severe receptive specific language impairment (SLI) were studied in 58 affected children (ratio of boys to girls 2:1) at the Department of Child Life and Health, University of Edinburgh, Scotland. When tested at a mean of 6 years after school entry, patients diagnosed with SLI had a normal mean non-verbal IQ, but one third had a persistent SLI, only 2 (3%) having attained normal scores on language tests. One third of siblings, previously considered normal, had abnormal language scores, and most family members had impaired phonological auditory memory, as measured by non-word repetition. A language learning difficulty occurred in 69% of first-degree relatives. Severe receptive SLI is almost always associated with severe reduction in expressive language skills. (Clark A, O'Hare A, Watson J et al. Severe receptive language disorder in childhood – familial aspects and long-term outcomes: results from a Scottish study. *Arch Dis Child* July 2007;92:614-619). (Respond: Professor Anne O'Hare, Department of Child Life and Health, 20 Sylvan Place, Edinburgh EH9 1UW, UK).

COMMENT. Receptive specific language impairment is characterized by difficulty in acquiring language comprehension despite normal non-verbal intelligence. Receptive language disorder has a poor prognosis and rarely resolves. Siblings and other family members are at increased risk of language impairments. Early diagnosis and intervention are recommended.

A follow-up study in later adult life, also at the University of Edinburgh, found that children with severe receptive developmental language disorder, reassessed in their mid-thirties, had deficits in verbal short-term memory and phonological processing, with social adaptation difficulties and increased risk of psychiatric disorder (Clegg J et al. *J Child Psychol Psychiatry* 2005;46:128-149).

Synthetic speech in treatment of language delay. Tallal P, at Rutgers University, and Merzenich MM et al, at University of California, San Francisco, reported remarkable and significant improvements in receptive speech and language comprehension in language-learning impaired children following training with acoustically modified speech stimuli. (Tallal P et al. *Science* 1996;271:81-84). Brief, rapidly changing components of speech were prolonged and emphasized, and coupled with adaptive training exercises. When presented in slower forms and rates, stimuli were correctly perceived and receptive language improved. (Merzenich MM et al. *Science* 1996;271:77-81). Compared to natural speech training, acoustically modified speech training was significantly more effective.

SEIZURE DISORDERS

EPILEPSY AND SPINOCEREBELLAR ATAXIA

A large consanguineous family from Saudi Arabia with 4 affected children presenting with an autosomal recessive ataxia, generalized tonic-clonic epilepsy and mental retardation is reported from the Institut de Genetique, Universite Louis Pasteur, Illkirch, France; Division of Pediatric Neurology, King Saud University, Riyadh, Saudi Arabia; and other centers. None of the 4 had myoclonus or mental deterioration. MRI of one patient revealed