

The Preserved Speech variant

Rett Syndrome is a neurodevelopmental disorder that mainly affects females. Nearly all females with Rett syndrome are lacking or are severely impaired in communication and receptive and expressive language. About 3-5 % belong to the preserved speech variant (PSV) that either preserve or recover the ability to speak to some degree. Girls with PSV have many of the same clinical features as females with classical Rett syndrome but with a more benign course. Some girls with PSV have been described as speaking in short phrases, often out of context, pronounced in the second and third person, and in an echolalic manner. Others have been reported to display more functional language use. The present paper will review the status quo of PSV, both the typical trajectory of speech, and the variation within the group. The paper includes three case profiles with different courses of development, level of abilities, and pragmatic language use, to further broaden the clinical spectrum of PSV. Girl 1 has a vocabulary of 15 words. She uses two of the words frequently and repeatedly, as verbal stereotypies. The speech of Girl 2 appears associative rather than referential. Her utterances appear in ritualized dialogues initiated by a partner who facilitates her speech by beginning utterances that she finishes, and as spontaneous responses to words said by other but without clear meaning. Girl 3 participates in longer conversations, she can read, write and do simple math. The implications of the review and the three different developmental courses are discussed.

References:

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