International Congress Series 1254 (2003) 199–205

Five children—vignettes of language disorders

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Abstract

The pediatric Otolaryngologist cares for children who have abnormal language as a primary or secondary deficiency. Five children, each with a different form of language disorder, are presented. These are children with specific language impairment (SLI) expressive, pervasive developmental delay (PPD, autism), expressive language delay associated with severe to profound hearing loss early in life, language delay secondary to a moderate to severe hearing loss diagnosed late and not cared for, and language delay secondary delay secondary to social deprivation and otitis media (OM) with effusion.

Keywords: Language delay; Language abnormality; Autism; Pervasive developmental delay; Social depravation; Cochlear implant; Otitis media with effusion; Delayed diagnosis; Language evaluation

1. Introduction

Language, a biological function, is a critical aspect of pediatric otolaryngology, both as a disease entity and as the outcome measure of many of the interventions we undertake. The following vignettes, all real patients in disguise, will show the manner in which these children may present to the pediatric Otolaryngologist and how they may be evaluated. Each child needs, in addition to the usual patient evaluation, a family history that explores speech and language disorders, an accurate hearing test, and a screen of language function. An instrument, which can objectively screen the patient's language level, is essential so that the practitioner can entertain a diagnosis and obtain the needed definitive evolutions and interventions. Coplan's Early Language

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doi:10.1016/S0531-5131(03)01131-2

Millstone Scale (ELM) is one of many such instruments and will be used in each case [1–3].

1.1. Pervasive developmental delay (autism)

This boy was first seen at 14 months of age because of a history of otitis media (OM). The history was unremarkable, both parents were professionals and the genetic history was negative for speech, language, and psychiatric disorders. The physical examination was within normal limits (WNL) and the pure tone audiogram showed normal hearing. The ELM (Fig. 1) was within age level but one would have expected somewhat better from a child of this social economic class.

The child returned at 26 months of age because he stopped speaking. He did not pay attention to others, and was almost constantly in motion. Physical examination and hearing were WNL. The ELM showed regression in both receptive and expressive language functions (Fig. 2). Based primarily on regression of language function with normal hearing, the tentative diagnoses of pervasive developmental delay (PPD) autism was made and confirmed by the pediatric neurologist. The child was placed in a therapy group and exposed to American Sign Language. Presently, he communicates primarily expressively with sign and uses both sign and aural language receptively.

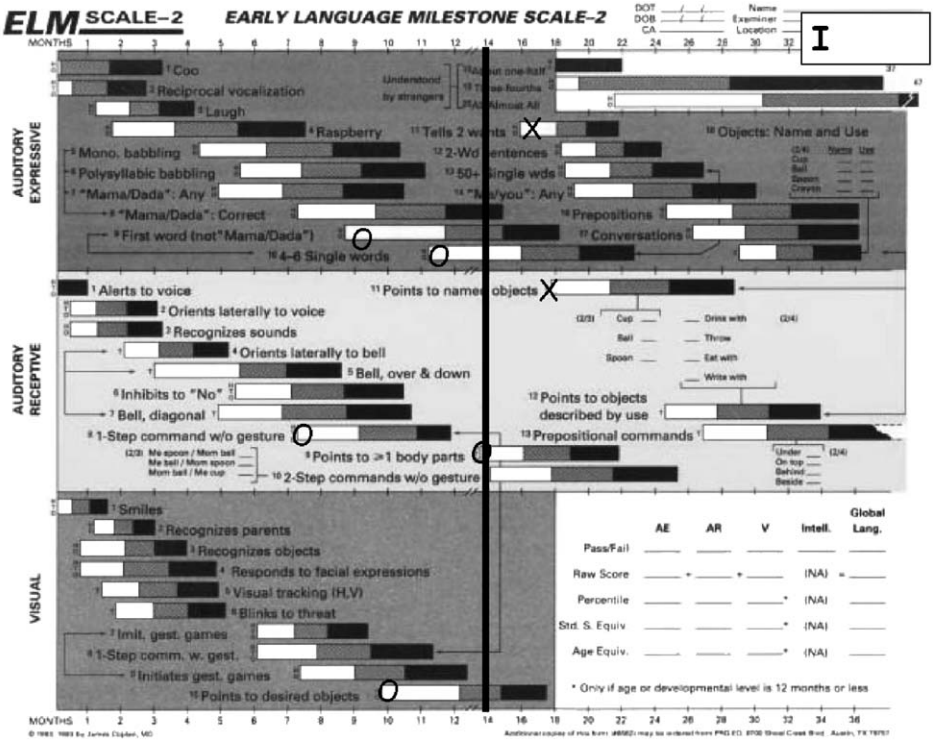

Fig. 1. ELM of child I at 14 months showing age level function.

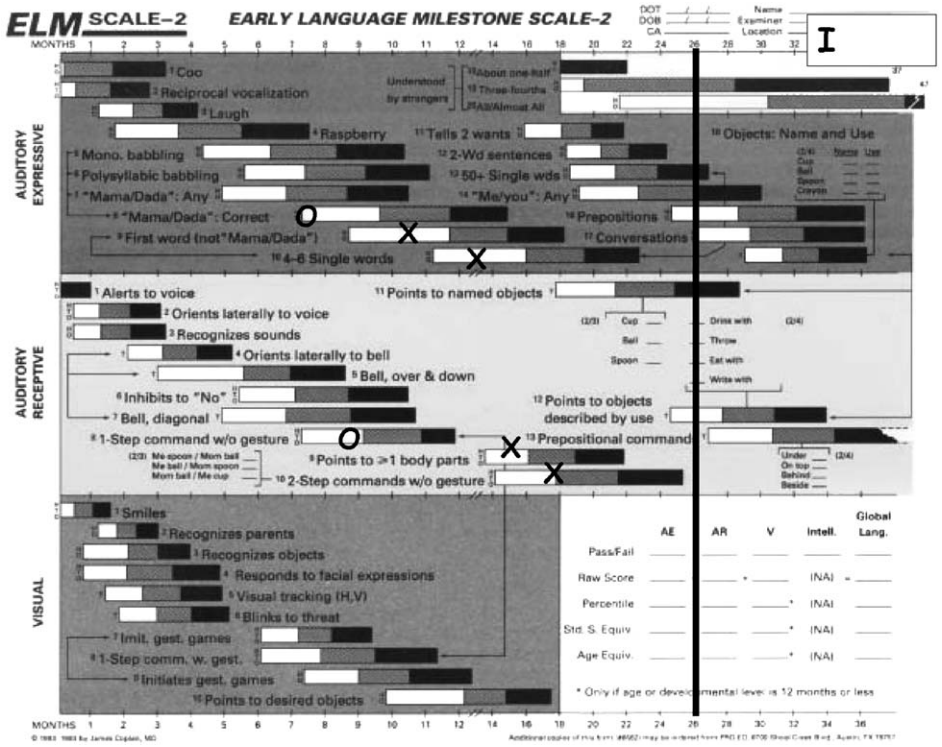

Fig. 2. ELM of child I at 26 months showing regression of language when compared to the ELM of Fig. 1.

1.2. Expressive language delay associated with conductive hearing loss early in life and social deprivation

This boy was seen at 10 months of age because of lack of any speech and recurrent otitis media. The past medical history revealed that he had been adopted from North Korea. His adoptive parents are both physicians and were born in Korea. He was malnourished and small for his age at the time of the adoption. The boy was small, quiet, and shy but not withdrawn. He had bilateral otitis media with effusion (OME). His hearing was assessed by visual reinforced audiometry (VRA) and he had a 25 dB conductive loss in both ears in the speech frequencies. The ELM showed that his receptive language was better than his expressive language and expressive language was below age level (Fig. 3).

Tympanostomy tubes were inserted and the hearing became normal. He was cared for in a bilingual home by his grandmother and parents and had speech and language therapy. At 36 months, he has normal hearing, is very playful, and exceeds age level English and is fluent in Korean.

This child is representative of the interaction of social deprivation—9 months in a North Korean orphanage with malnutrition and the hearing loss of OME. At the

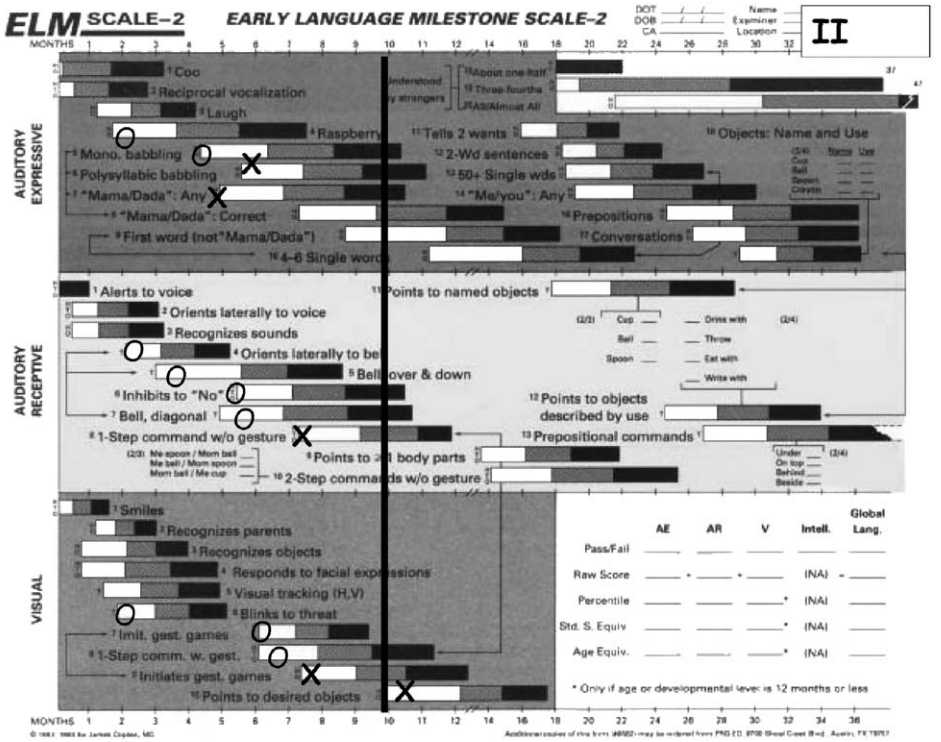

Fig. 3. ELM of child II at 10 months showing delayed expressive language function.

age of 10 months, he received restoration of hearing, a socially and linguistically enriched environment, and special help, all of which contributed to a satisfactory outcome.

1.3. Language delay secondary to a moderate to severe hearing loss and delayed diagnosis

This 30-month-old child was seen because she did not talk as well the other children in her playgroup. Their pediatrician reassured her parents that there was nothing to worry about and that she would outgrow this. The child's grandmother felt that she was not hearing well and finally persuaded her parents to seek a second opinion. The past history was unremarkable and there was neither history of hearing loss in the family nor was there any suggestion of consanguinity. The physical examination was WNL. The pure tone audiogram showed a bilateral slopping sensory hearing loss with a pure tone average (500, 1k, 2k, and 4k) of 60 dB. The ELM found both the expressive and receptive language levels to be at 15 months, giving a language quotient of ~ 50% (Fig. 4).

Hearing aids, speech, and language therapy were recommended. The parents did not avail themselves of these interventions for they thought that the child could hear and did

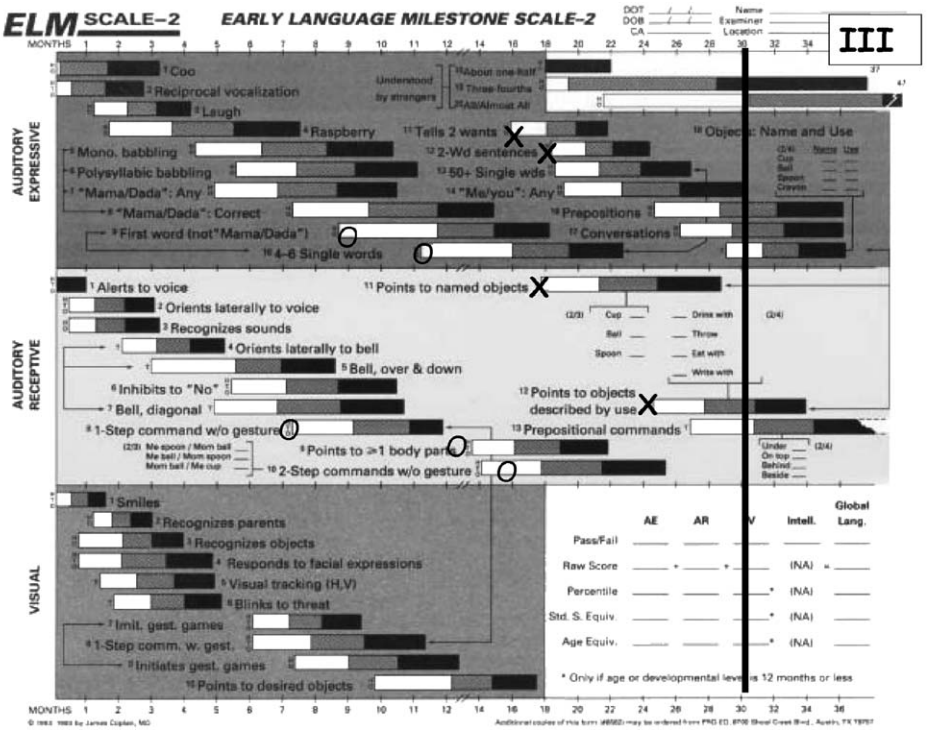

Fig. 4. ELM of child III at 30 months showing delayed expressive and receptive language functions.

not need a stigmatizing hearing aide. The child was seen again at age 5, the hearing was the same and the child's language quotient remained at ~ 50%. There is little likelihood, that at age 5 years, a hearing aid and therapy will be effective in restoring normal language function [4].

1.4. Specific Language Impairment (SLI) expressive

This child was seen first at 34 months, referred from his preschool, because he was not talking. The medical history was unremarkable except for the genetic history. His father, a stonemason, did not talk until 3 years of age and had speech and language therapy in school. A maternal aunt was a late talker. The physical examination was WNL as was the audiogram. ELM shows receptive at age level and expressive at the 22-month level for this 34-month-old child (Fig. 5). The diagnosis of specific language impairment (SLI) was made and the child entered into a speech and language program. One of the other four siblings was found to have SLI.

At 7 years of age, the patient is in a regular school with language therapy. The child's expressive language is almost at age level and his receptive language is much better than would be expected at 7 years.

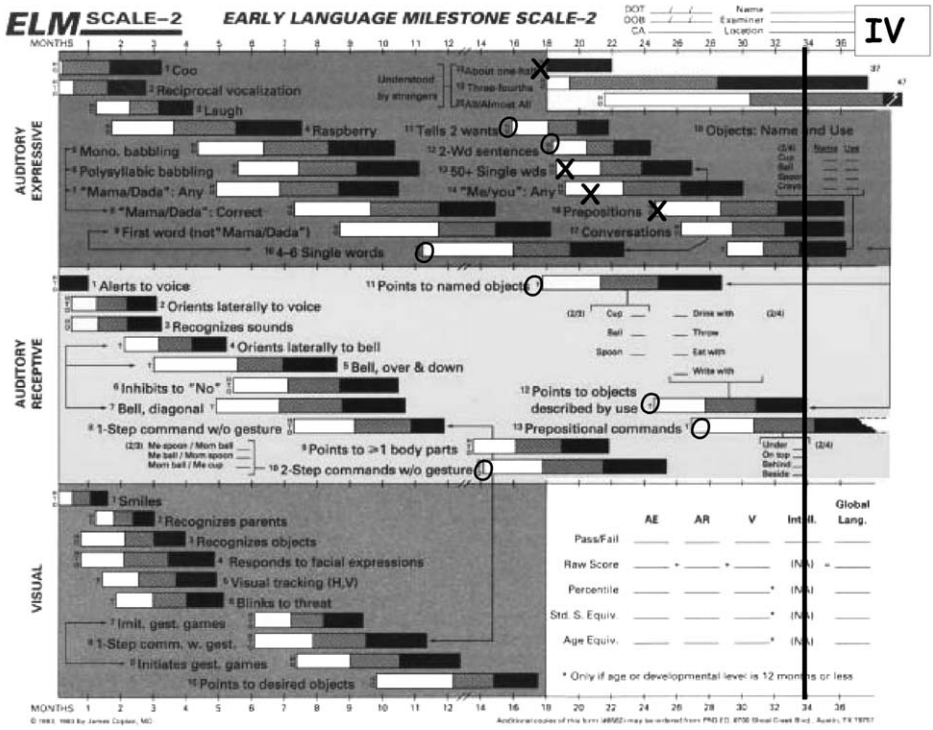

Fig. 5. ELM of child IV at 34 months showing delay of expressive and receptive language function.

1.5. Language delay with early onset profound hearing loss

The patient was seen at 9 months because of a suspected hearing loss. Her pre- and perinatal histories were unremarkable and she passed a newborn hearing screening. Her parents noted that at 6 months she stopped rolling over, was not paying attention to sound but enjoyed visual objects. She would smile when a parent entered her view. The medical history is genetically significant. Both sides of the family have northern Mediterranean origins and the parents are second cousins. There is a history of hearing loss, static, and progressive on one side of the family. Her older brother has surgically proven perilymphatic fistulae and early onset loss progressive hearing loss that was not diagnosed until age 3.

Physical examination was unremarkable other than the patient was a very quiet baby. Auditory function is assessed with VRA, otoacoustic emissions (OAEs) and auditory brain stem response (ABR) showed no hearing in one ear and a severe loss in the other. Vestibular tests were positive for fistulae. The ELM showed delayed expressive and receptive language and normal visual skills (Fig. 6).

She was diagnosed as a Connexin (GJB2) compound heterozygote 30delG/167delT. She has surgically proven perilymphatic fistulae that were repaired a number of times,

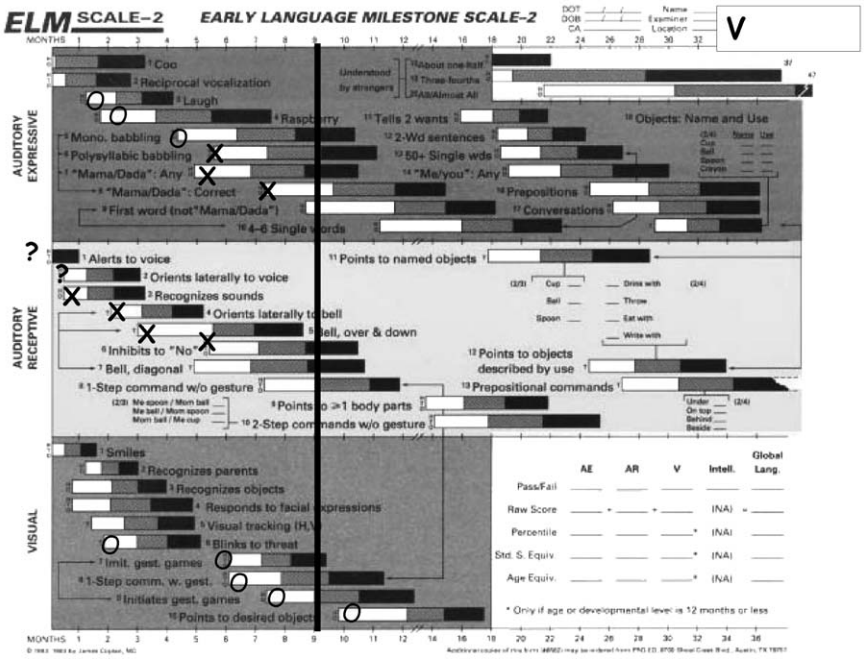

Fig. 6. ELM of child V at 9 months showing delay of expressive and receptive language function.

amplification at 9 months, and extensive speech and language therapy. At 6 years of age, her nonhearing ear underwent cochlear implantation. At 9 years old, she is an honor student in a normal school and is bilingual.

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