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WHAT EVERY EDUCATOR SHOULD KNOW ABOUT LANDAU-KLEFFNER SYNDROME

Landau-Kleffner syndrome is a rare and unique syndrome that is most often characterised by difficulty in receptive or expressive language ability, abnormal electroencephalograms (EEG's), and seizures. It is important for educators to understand this distinct syndrome because children with Landau-Kleffner syndrome are often misdiagnosed. In this article we synthesise the literature on the primary and secondary characteristics of children with Landau-Kleffner syndrome (eg. behaviour problems). A framework for intervention techniques is then provided based on the literature on appropriate techniques for children with language difficulties. Interventions discussed include using predictable language, creating the need to communicate, and using alternative and augmentative communication techniques.

Mrs. Smith is a first-grade teacher who is considering writing a referral for one of her students. This child has recently experienced a drastic change in his behaviour. He refuses to participate in class activities and does not follow instructions. Mrs. Smith has tried talking to him about his behaviour, but he usually does not answer her questions. Although his hearing was tested and found to be normal, he acts as though he cannot hear Mrs. Smith when she is talking to him. When he does answer a question, he mumbles something that is incomprehensible. In addition, his academic skills have decreased and he has developed behaviour problems. His parents report the same problems at home and interpret his behaviour as "just plain stubborn." There have been no major changes in the family environment. Mrs. Smith is aware of these facts and is astonished by her student's completely uncharacteristic behaviour. He has always been one of the most social, well-behaved, and self-motivated students in her class. Why has this suddenly changed? Although there can be several reasons for this child's drastic change in behaviour, one possibility is that her student has a syndrome that is extremely rare and completely unknown to most educators Landau-Kleffner syndrome.

Landau-Kleffner syndrome is particularly important for teachers to understand given the complexity and rarity of this disorder and the lack of information currently available on the characteristics of students with this syndrome and appropriate intervention strategies. Developing a knowledge base in order to identify the characteristics of Landau-Kleffner syndrome is important for educators because children with this syndrome arc often misdiagnosed as having other disorders that have some similar characteristics, such as autism, epilepsy, or Rett syndrome (National Organisation for Rare Disorders, 1995). The first purpose of this article is to synthesise the published research on the primary and secondary characteristics of children with Landau-Kleffner syndrome. The second purpose is to provide teachers with a framework for designing interventions for students with the syndrome based on their unusual needs and characteristics.

Primary Characteristics

Acquired aphasia with convulsive disorder in children, also known as Landau-Kleffner syndrome, is both a rare and idiosyncratic syndrome. It was first cited in a research report that included case studies of six children, ages 5 to 15 years, who had developed aphasia (ie. the loss or deterioration of speech comprehension and production abilities; Landau & Kleffner, 1957). Children who were participants in this study also had convulsions that accompanied, preceded, or followed the aphasia. All of the children studied had electroencephalographic abnormalities. The onset of the disorder was sudden in some cases and gradual in others. Language improvement occurred in all six cases. Interestingly, two of the six children had a complete recovery of their previously acquired language skills without any systematic or planned speech therapy. Children in this study were described as having a newly discovered and very complex disorder named after the researchers-Landau-Kleffner syndrome.

Since 1957, at least 160 other case studies involving children with Landau-Kleffner syndrome have been reported (National Organisation for Rare Disorders, 1995; Paquier, Van Dongen, & Loonen, 1992). The available research has included children with symptoms similar to those first studied by Landau and Kleffner (eg. Cooper & Ferry, 1978; Maccario, Hefferen, Keblusek, & Lipinski, 1982; White & Sreenivasan, 1987). The research has overwhelmingly indicated that children with Landau-Kleffner syndrome have abnormal EEG's and a loss of receptive and expressive language skills or abilities. However, research has also documented that not all of the children studied experienced observable convulsions (eg., Jordan, 1980; Maccario et al., 1982). According to a review of the cases cited in the literature, approximately 80% of children with Landau-Kleffner syndrome experienced seizures (Paquier et al., 1992). Medication is typically very effective in treating the seizures manifested by children with Landau-Kleffner syndrome. Thus, from an extensive number of case studies, the primary characteristics of children with Landau-Kleffner syndrome are abnormal EEG, loss of language, and possible seizures.

Two additional factors to consider when determining who may have Landau-Kleffner syndrome are gender and age at onset. Case study research has found that males are two times as likely to have Landau-Kleffner syndrome as females (National Organisation for Rare Disorders, 1995). Age at onset of Landau-Kleffner syndrome is typically between 3 and 7 years (National Organisation for Rare Disorders, 1995). Determining the age at onset is very important because it is associated with both the severity of the manifestation of the syndrome and the long-term outcome (Bishop, 1985). Bishop studied the relationship between age at onset and severity, of disorder in 61 children with Landau-Kleffner syndrome who were followed to the age of 12 years. Early onset of the disorder was significantly correlated with a more severe language disorder and a poor long-term outcome (eg., less likelihood of recovery). More detailed analysis of the 33 children whose onset occurred between birth and age 5 showed that 1 child had normal language, 1 had mild articulation problems, 8 had mild language problems in word finding and speech comprehension, 8 had moderate language difficulties, and 15 (45%) had severe language disorders with little ability to comprehend spoken language. These findings illustrate the importance of early intervention for children with Landau-Kleffner syndrome.

Language Ability and Children with Landau-Kleffner Syndrome

Loss of language by students with Landau-Kleffner syndrome may be the most salient characteristic noted by teachers. Researchers have described the first sign related to the language loss in children with Landau-Kleffner syndrome as "word deafness" (Paquier et al., 1992, p. 354). Children with Landau-Kleffner syndrome also tend to have difficulty using language to express themselves (eg. Cooper & Ferry, 1978; Deonna, Fletcher, & Voumard, 1982; Maccario et al., 1982; Paquier et al, 1992; White & Sreenivasan, 1987). Children with Landau-Kleffner syndrome may have normal voice quality, and inflection (Worster-Drought, 1971) or have a high-pitched voice with abnormal inflection similar to that of children who are deaf (Landau & Kleffner, 1957). Children with Landau-Kleffner syndrome may also imitate another person's speech (ie. echolalia; Maccario et al.) or persevere on one syllable in their speech or one letter in their writing (Landau & Kleffner). Children with this syndrome also have articulation problems that range from mild to severe (Jordan, 1980; Maccario et al.), with some children demonstrating unusually long pauses between syllables when pronouncing single words (Deonna et al.). Other speech-language problems that may be present in children with Landau-Kleffner syndrome include dysnomia (eg. say "hammer" for shovel), semantic paraphasia (eg. say "cut" for scissors), and reverse compounds (eg. say "lightstop" for stoplight, Maccario et al.). In noisy situations, children with Landau-Kleffner syndrome have even greater problems discriminating speech (Jordan, 1980).

Some children with Landau-Kleffner syndrome have described their own expressive and receptive language difficulties after they have recovered their language abilities. In the original study conducted by Landau and Kleffner (1957), one child reported that when people were talking to him, it sounded like jargon ("blah"). Another child from this same study reported that he understood what other people were saying but could not retrieve the words that he wanted to use to respond. In another case study report, Jordan (1980) observed that one child with Landau-Kleffner syndrome responded to a question such as "How are you?" with the correct response to the question "Where do you live?" Language comprehension problems were also systematically studied in other research that found that children with Landau-Kleffner syndrome as a group have deviant language patterns and rigidly use word order and sequence to interpret sentences rather than use the grammatical structure of the sentence (Bishop, 1982). Bishop also documented that the inaccurate language interpretation shown by children with Landau-Kleffner syndrome was similar to that of children who are deaf.

To illustrate further the types of errors made by children with Landau-Kleffner syndrome, take the following sentence: "The book on the shelf is red." For this sentence, children with Landau-Kleffner syndrome tend to state that the shelf is red instead of the book on the shelf. Thus, rather than interpret how a complement refers to the subject of a sentence, they simply link the complement to the nearest noun (ie. reliance on word order). Last, the language abilities of children with Landau-Kleffner syndrome have been documented in the literature as inconsistent (eg. Paquier et al., 1992). That is, language gains made by children with Landau-Kleffner syndrome may be lost at any time, and language deficits may improve without extensive interventions.

Secondary Characteristics

An important factor to consider when reviewing the following secondary characteristics associated with Landau-Kleffner syndrome is that they vary in both number and severity. Some children may experience all of the following characteristics, and some may experience only one or two. Children may manifest a particular characteristic for only a couple of days, for weeks, for months, or possibly throughout their entire lives.

Aptitude. Most children with Landau-Kleffner syndrome are reported to have low-average to above-average intelligence (Landau & Kleffner, 1957; Maccario et al., 1982). Children with the syndrome are also reported to have strong visual learning skills (eg. Cooper & Ferry, 1978; Jordan, 1980; Landau & Kleffner, 1957; Maccario et al.). These visual strengths include visual memory, closure, association, recognition, comprehension, identification of details, perception, and analysis. Some children with Landau-Kleffner syndrome communicate by reading and writing in addition to or instead of communicating orally (Cooper & Ferry; Jordan). Many children with the syndrome have strengths in other forms of nonverbal communication, such as sign language or gestures (Maccario et al.). The nonverbal skills of children with Landau-Kleffner syndrome range from pointing to objects and pictures to a complete comprehension of sign language.

Although students with Landau-Kleffner have some relative strengths, there are also some needs areas. Research shows that while students with the syndrome may comprehend well visually, many have difficulty comprehending auditorally (Jordan, 1980). Some studies have documented that children with Landau-Kleffner syndrome have auditory memory (Cooper & Ferry, 1978; Jordan; Landau & Kleffner, 1957; Maccario et al., 1982) and auditory processing problems (Cooper & Ferry; Maccario et al.).

Social/Behavioural. Children with Landau-Kleffner syndrome have also been reported to be at risk for developing social and behavioural problems. The National Organization for Rare Disorders (1995) reported that 70% of children with the syndrome evidence some behaviour problems, including aggression, eating and sleeping problems, and hyperactivity. However, case study research has generally documented that children with Landau-Kleffner are described as friendly (Jordan, 1980; Landau & Kleffner, 1957).

Behaviour problems, such as "stubbornness," difficulty, following lengthy directions, and refusal to participate in class activities, may relate directly to problems in understanding language and not too general noncompliance. However, if educators are not able to distinguish a language problem from a behaviour problem, they may interpret children's behaviour as intentional and purposeful. Possibly because of their difficulty understanding spoken language, children with Landau-Kleffner syndrome have been reported to have difficulty following more lengthy verbal directions (Cooper & Ferry, 1978; Jordan, 1980; National Organization for Rare Disorders, 1995) but to be able to follow simple commands when accompanied by gestures (Maccario et al., 1982).

To summarise the behavioural research, as a group children with Landau-Kleffner syndrome have varying degrees of the following behaviour problems that may be present in the classroom: hyperactivity, aggressive behaviour, stubbornness, and difficulty following lengthy oral directions (Cooper & Ferry, 1978; Jordan, 1980; National Organization for Rare Disorders, 1995; Paquier et al., 1992; Sawhney, Suresh, Dhand, & Chopra, 1988; White & Sreenivasan, 1987).

Framework for Interventions

There is little research concerning specific educational interventions for children with Landau-Kleffner syndrome. According to the first report by Landau and Kleffner in 1957, it appears that specific classroom interventions may not be necessary, because some children show improved language skills regardless of whether

they receive medication or specialised intervention. Because it is not possible to know who will improve without intervention and who will not, intervention is extremely important. Thus far, no research is available that compares the success of different language intervention strategies for children with Landau-Kleffner syndrome. In fact, no reports of specific language interventions have been documented for use with children with Landau-Kleffner (Paquier et al., 1992).

One study found that attempts to elicit spoken communication and improve receptive language skills in children with Landau-Kleffner syndrome were unsuccessful in 42% of the cases reviewed (Cooper & Ferry, 1978). If receptive and expressive language skills do not improve for children with Landau-Kleffner syndrome, then teachers and professionals need to make decisions regarding other modes of communication that can be used, in addition to language interventions, so that the child can communicate (Cooper & Ferry). Deciphering which intervention is most effective with a particular child will take trial-and-error testing, especially until more research is conducted in this area. The following intervention strategies are appropriate for children experiencing difficulties in language and can serve as a framework for selecting interventions based on the individual characteristics of a child with Landau-Kleffner syndrome (eg. strong visual memory). However, it is important to note that there is not yet any empirical validation for the use of these strategies for children with Landau-Kleffner syndrome. Therefore, interventions must be closely monitored to ensure an appropriate match between an individual child and a particular intervention strategy.

Use predictable language. Repetition and redundancy are extremely important in teaching children with language problems. Cook, Tessier, and Klein (1996) specified that "certain key words and phrases should be used repeatedly to make significant the events of the day" (p. 338). Consistently pairing words with certain daily events helps establish verbal routines for the child.

Create the need to communicate. Encouraging children to use the language they have can be done by creating the need for children to use language. Ostrosky and Kaiser (1991) have recommended a set of strategies that can be used by teachers to create the need to communicate:

- Place desirable objects within view, but out of the child's reach. This will require the child to request the object. Reinforce the child's request if he or she requests the object in sign language or orally, whichever is most appropriate.

- Provide inadequate portions of desired materials. Reinforce the child's request for more. The teacher can model or prompt more elaborate language when appropriate.

- Present two or more materials or activities from which the children may choose. This information can be gathered by observing what the children like to play with or by asking their parents.

- Reinforce the desired behaviour, whether it is spoken or unspoken communication.

- Create situations in which the child is likely to need some help. For example, putting toys in a container that the child will need help opening will require that the child communicate with someone to get to the toys. However, care must be taken with this strategy to keep the frustration level of the child at a minimum.

- Purposely leave out an object that a child needs to complete an activity. For example, provide the child with paper and paint but don't provide the paintbrush.

- Set up a silly situation that does not meet the student's expectations. For example, while playing, the teacher could put on a child's shoe, encouraging the child to laugh and communicate that something is wrong with the situation.

Shape communicative responding. Sometimes the desired behaviour must be shaped. Cook et al. (1996) have defined five structured training steps in the shaping of specific communicative behaviours:

1. "Create the need or opportunity to communicate."

2. "Pause and wait (for communication)."

3. "Provide a natural cue."

4. "Use prompts and assistance."

5. "Comply with the communicative re-quest" (pp. 341-342).

Use alternative/augmentative communication. For some children, opportunities to communicate must include nonspeech responses. Thus far, except for sign language, there has been no research on the use of alternative or augmentative communication systems for children with Landau-Kleffner syndrome. Therefore, teachers should choose an alternative/augmentative system that best meets the need of their student. For all students, alternative/augmentative systems should be used in addition to, not instead of-speech. In addition, speech should be augmented with physical gestures and facial expressions to help children make meaning of the words.

Some research has been done on the use of sign language with children with Landau-Kleffner syndrome. Bishop (1982) found that children with the syndrome had deficits in language stemming primarily from lack of understanding of sentence structure. Bishop also found that children who are deaf have a pattern of performance strikingly similar to that of children with Landau-Kleffner syndrome. One sign system is the Paget-Gorman Sign System. This "artificially devised sign system uses the same word order as spoken English and includes signs for function words and inflections..." (Bishop, 1982, p. 3). Bishop found that children with Landau-Kleffner syndrome have comprehension deficits when using this type of sign system.

If it is true that the comprehension problems of children with Landau-Kleffner syndrome are due primarily to misinterpretation of grammatical structure (Bishop, 1985), a sign system that might be effective is American Sign Language (ASL). Each sign represents a concept, not a word, and has its own syntax and grammar. It is not a word-for-word representation of the English language (Flodin, 1994). For example, instead of signing the words "I want something to eat" (which consists of five separate signs), a child would sign only one word, "eat." Using sign language along with spoken instruction can help increase the child's communication skills. This may in turn increase his or her academic achievement and behaviour competence.

For children who are unable to speak, sign, or write, an alternative communication system can be used. Again, the alternative system should be combined with speech. One alternative communication device is a communication board. Communication boards may be low tech and inexpensive, such as those made by pasting pictures of various objects to construction paper and laminating, or they may be high tech, using computerised systems that "talk" for the child. In either of these options, the child is then required only to point to a picture to communicate a need or to answer a simple question.

Use visual supports. The use of visual supports should be embedded in the child's entire educational program. These may include objects, models, pictures, demonstrations, sequenced information, outlines, or gestures, anything that supports the student's visual strengths (Dairypie & Ruble, 1995). Following are some examples of visual supports.

1. **Schedules:** Photograph various objects that are representative of different activities throughout the day. Laminate the pictures and secure a piece of velcro under the picture. Next, laminate an arrow and secure the other side of velcro to it. The child can then move the arrow to each activity to help ease transitions. This strategy communicates to the child what is going to happen next. Depending on the pictures, the schedule lets the child know what he or she needs to do, the materials needed, and where to go for the activity. It may also be a good idea to have a small version of the laminated schedule on the student's desk and have him or her check off different activities and items that will be needed for each activity throughout the day (Stormont-Spurgin, 1997).

2. **Rules:** Take photographs of students following the rules of the classroom or performing appropriate behaviour. For example, during large-group instruction, show the child a picture of himself or herself or a peer model sitting still and looking at the teacher. The picture communicates to the child what he or she should be doing in that setting.

3. **Desired behaviour and consequences:** Post a picture of a desired behaviour and couple it with a picture of the reinforcer. This pairing also communicates to the child what to do and what will happen when he or she does it. This can be used as a contingency contract to change a problem behaviour.

4. **Use behavioural supports:** Teaching a child with behaviour problems is always a difficult task. When a child has Landau-Kleffner syndrome and the accompanying language deficits, teaching can be even more

difficult. Dalryple and Ruble (1995) recommended the following proactive behavioural supports for working with children with the syndrome:

a. Tell the child what to do instead of what not to do (remain positive).

b. If a child does not want to do a task, analyze the possible reasons from the perspective of the child (task/ situational analysis).

c. Use individual motivational activities and materials.

d. Make rules and routines very clear. Make sure the child understands them by teaching visually and concretely and using extensive repetition.

e. Teach effective communication and social interaction techniques.

f. Teach strategies for dealing with anxiety.

Because children with Landau-Kleffner syndrome have been described as friendly and well liked, and because of their strong imitation skills, peer models could also be used in teaching new skills or other desired behaviours. Negative behaviours, such as aggressiveness, should be dealt with consistently. Consistency, structure, and routines are extremely important in teaching skills to all children and particularly important when teaching children with Landau-Kleffner syndrome.

Modify the listening environment. Some researchers have suggested ways to modify the environment to improve the receptive language abilities of children with Landau-Kleffner syndrome. Tharpe, Johnson, and Glasscock (1991) suggested changing the interference of background noise in classrooms by improving the speech signal-to-noise ratio (SNR). Environmental modifications could include acoustic treatments of classrooms or use of personal frequency modulation (FM) systems.

Summary

To date, we have not located an article that has reviewed the characteristics of children with Landau-Kleffner syndrome for the purpose of better preparing teachers to teach such students. One purpose of this paper was to review the characteristics of children with the syndrome. A second was to provide a framework for linking the individual characteristics of children with Landau-Kleffner syndrome to help educators design appropriate interventions for such children. Educators should keep data on the effectiveness of selected interventions because each child with Landau-Kleffner syndrome, and each child in general, is different and requires individualised interventions.

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