

Critical Review:
Evidence for Residual Long-Term Speech Deficits Following Transient Cerebellar Mutism in Childhood

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This critical review examines the available evidence for residual long-term speech deficits following transient cerebellar mutism in childhood. Study designs include: parental survey, retrospective chart review, between group design, and case studies (3). Overall, the research supports the presence of residual speech deficits (articulation, fluency, phonology, rate of speech, and dysarthria) in many individuals who underwent surgery and recovered from mutism years prior. The results of the present review should be interpreted with consideration of the inherent limitations of methodology used in the reviewed studies.

Introduction

The cerebellum is involved in controlling the quality of speech production by altering and regulating the movements of the speech musculature after they have been planned by the central nervous system (Gordon, 1996). Speech movements become disjointed and disconnected if the muscles are not accurately started and stopped as has been observed in individuals with cerebellar lesions (Gordon, 1996). Cerebellar damage may be caused by stroke, tumour resection, alcohol abuse, head injury, or exposure to chemicals such as toluene and phenytoin. Cerebellar lesions may result in any type of dysarthria affecting the ability to coordinate the movements of the tongue, lips, and palate, as well as the synchronization of respiration and phonation (Gordon, 1996). The most typical speech pattern resulting from cerebellar lesions is that of ataxic dysarthria which is characterized by imprecise consonants, excess and equal stress, irregular articulation breakdown, vowel distortion, harsh voice, phoneme and interval prolongation, mono pitch and loudness, and slow rate (Huber, Bradley, Spiegler & Dennis, 2006).

In addition to the characteristics of ataxic dysarthria stated above, some individuals develop transient cerebellar mutism, also known as posterior fossa syndrome or cerebellar syndrome, following surgery to remove cerebellar tumours. This transient mutism, which is a state of speechlessness in a conscious patient, occurs in 7.5% of patients following tumour resection and is more common in children than adults (Huber et al., 2006). The posterior fossa is the most common site for cerebellar tumours in children and the cerebellar astrocytoma and medulloblastoma tumours together account for approximately one third of all childhood tumours (Huber et al., 2006).

In children who develop transient cerebellar mutism following tumour resection, well-preserved speech is

observed for 24 hours up to 6 days, after which time mutism begins and can last anywhere from several days to several months, or even years (Gordon, 1996; Huber et al., 2006; van Dongen, Catsman-Berrevoets, & van Mourik, 1994; Steinbok, Cochrane, Perrin, & Price, 2003; Doxey, Bruce, Sklar, Swift & Shapiro, 1999). As speech begins to re-emerge following the period of mutism, children often present with the characteristics of ataxic dysarthria discussed above. Therefore, the term 'mutism with subsequent dysarthria' has been used in the literature to refer to the process of the recovering cerebellar mechanism (Huber et al., 2006). The nature of cerebellar recovery following tumour resection is not fully understood. Some researchers describe the mutism as resolving completely into normal speech (Dietze & Micle, 1990; Pollack, Polinko, Albright, Towbin, & Fitz, 1995; Van Calenbergh, Van De Laar, Plets, Goffin & Casaer, 1995; Cochrane, Gustavsson, Poskitt, Steinbok & Kestle, 1994). Other researchers suggest that speech never fully recovers (Huber et al., 2006; Huber-Okraïnec, Dennis, Bradley, & Spiegler, 2001; Steinbok, Cochrane, Perrin & Price, 2003). This discrepancy in the literature has been noted by researchers and some have gone so far as to suggest that "One might reasonably conclude from a review of the literature that 'cerebellar mutism' is a distressing, but transient and ultimately benign problem" (Steinbok, Cochrane, Perrin & Price, 2003, pg. 180).

Objectives

The primary objective of this paper is to critically evaluate the existing literature regarding what evidence there is for residual long-term speech deficits following transient cerebellar mutism in childhood. The second objective is to provide evidence-based recommendations for future research.

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Methods

Search Strategy

Computerized databases, including Commdis Dome, CINAHL, Pubmed, and Medline, were searched using the following search strategy:

((transient cerebellar mutism) OR (posterior fossa syndrome) OR (cerebellar syndrome) OR (mutism)) AND (dysarthria) AND ((tumour resection) OR (tumor resection)).

The reference lists of the articles found were also searched for relevant papers.

The search was limited to articles written in English.

Search Criteria

Studies selected for inclusion in this review were required to investigate speech characteristics, involve individuals who had cerebellar tumours resected in childhood, and who developed a transient period of mutism as a result of the cerebellar lesion. In order to fulfill the requirement of 'long-term' deficits, studies were also required to have participants who had received surgery at least 2 years prior.

Data Collection

Results of the literature search yielded the following study types: parental survey, retrospective chart review, between group design, and case studies (3).

Results

In their 2006 study Huber, Bradley, Speigler and Dennis investigated the presence of residual motor speech deficits in six survivors (mean survival years was 10.78) of childhood transient cerebellar mutism (TCM) after cerebellar tumour resection. These individuals were then compared to six individuals with cerebellar tumours who did not develop postoperative TCM and six healthy individuals in order to determine whether deficits were greater in those individuals who developed TCM than in the other two groups. Subjects were videotaped while providing a narrative in response to a picture book. Two speech-language pathologists (SLPs) independently analyzed the videotaped narratives to determine percent dysfluencies, rate of speech, and the presence of ataxic dysarthria using the Dysarthria Rating Scale. Results showed that individuals who developed TCM postoperatively were significantly more dysarthric and had slower rates of speech than either the healthy controls or the individuals who did not develop TCM postoperatively. The results also showed that patients who developed TCM following tumour resection were significantly more dysfluent than healthy controls, but were not significantly

different from patients who did not develop TCM following tumour resection.

The 2003 study by Steinbok, Cochrane, Perrin, and Price used parental reports to determine the long-term neurological and speech outcomes of seven patients who developed TCM following cerebellar tumour resection in childhood. Subjects were identified through a search of a hospital's database and medical records were reviewed and parents were contacted to determine their child's most recent speech and neurological status. Reports from parents indicated that at time of follow-up (between 2.5 and 13.1 years post-surgery) one child remained mute, one child's speech returned to normal, three children had speech that was reported to be slower than normal, and two children were reported to slur their speech.

Doxey, Bruce, Sklar, Swift, and Shapiro (1999) performed retrospective chart reviews to determine the reversibility of complications following tumour resection and TCM. The medical records of twenty individuals were reviewed for the development of postoperative neurological deficits. Results indicated that at time of follow-up (between 0.5 and 16 years post-surgery) seven individuals had 7th nerve palsy and four individuals had bulbar palsy. Although not described in the paper, these neurological deficits may lead to difficulties in speech due to weakness of the speech musculature.

In 1989, Hudson, Murdoch, and Ozanne looked at the presence of articulation, phonological, phonetic, and motor speech deficits in two individuals who had undergone surgery in childhood for the removal of a cerebellar tumour and developed TCM postoperatively. Subjects completed the Fisher-Logemann Test of Articulation Competence and the Khan and Lewis procedure was used to analyze the results phonetically. Participants also completed the Frenchay Dysarthria Assessment and provided an audio recorded connected speech sample which two independent SLPs used to determine the presence or absence of the ten most prominent features of ataxic dysarthria as defined by Darley, Aronson, and Brown (1969). The first individual, who was 6 years post-operative, was described as presenting with a number of inconsistent phoneme productions, the retention of phonological processes, a mixed ataxic-flaccid dysarthria, left facial palsy, lack of volume control, and five of the ten ataxic dysarthria characteristics were reported to be present including imprecise consonants, excess and equal stress, irregular articulatory breakdowns, prolonged phonemes, and slow rate. This individual was reported to be largely unintelligible. At four years

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and nine months post-surgery, the second individual was described as presenting with some phoneme prolongations, pitch breaks, variable pitch, lack of volume control, explosive onsets, and six of the ten ataxic dysarthria features including imprecise consonants, excess and equal stress, harsh voice, prolonged phonemes, prolonged intervals, and slow rate.

Di Cataldo, Dollo, Astuto, La Spina, Ippolito, and Papotto (2001) provided the case of two individuals who at 25 and 34 months after surgery for the removal of a cerebellar tumour were reported to have normal speech following TCM. No formal assessment was completed.

In 1996, Jones, Kirollos, and Van Hille described the case of an individual who underwent cerebellar tumour resection and developed TCM postoperatively. At a two year follow-up the individual was reported to demonstrate slurring and stuttering of speech, although no formal assessment was completed.

Discussion

Appraisal of the Results

When reviewing the results of this paper, it is important to consider issues with regard to subject selection, sample size, statistics, and methodology, as these factors may impact the strength of the evidence.

Subject Selection and Sample Size

The small sample size of the studies ranged from one to twenty subjects which compromises the generalizability of the findings.

Another issue of these studies is the lack of random sampling of participants. The participants in the study by Huber et al. (2006) were the same individuals that participated in a previous study and the methods of participant selection were not described in either paper. Therefore it is unknown whether the means of selection were valid and thus to what extent the results can be generalized. A number of authors (Steinbok et al., 2003; Doxey et al., 1999; Hudson et al., 1989) used hospital databases in order to identify potential subjects for their studies. This process may have inherent biases as individuals who seek services at a particular hospital may be of a certain ethnic background or have a certain socioeconomic status. However, this background information was not provided by the aforementioned authors and therefore what population the subjects are representative of is unknown, reducing generalizability.

Information regarding participant inclusion and exclusion criteria was not included in a number of studies (Huber et al., 2006; Steinbok et al., 2003; Doxey et al., 1999; Di Cataldo et al., 2001) which means that the medical history and presence of premorbid speech impairments of the subjects is unknown and therefore the effect of these pre-morbid conditions on the findings are unidentified. None of the papers reported whether the subjects had received any speech therapy and therefore it is unknown whether the results obtained can be generalized only to individuals who receive therapy or only to those who do not. Reporting and controlling for these variables would assist in ensuring that the results obtained were due to the effects of cerebellar tumour resection and TCM and not to premorbid medical issues or speech impairments.

Methodology

When interpreting the results of studies, it is important to consider limitations of the methodology, in particular prominent information that was not included in the research descriptions, statistics provided, and the type of study conducted (e.g. case study, chart review, experimental).

A number of researchers (Huber et al., 2006; Hudson et al., 1989; Doxey et al., 1999) failed to state how many times the SLPs were able to listen to speech samples, who administered the standardized tests and how, and how neurological deficits were diagnosed and by whom. This lack of information reduces the reproducibility and validity of the findings.

A number of researchers relied on subjective observations made by themselves in their case studies (Di Cataldo et al., 2001; Jones et al., 1996), or by parents through parental reports (Steinbok et al., 2003), to determine the presence of speech deficits. These studies do not provide empirical findings and therefore the results are less valid and reliable than experimental approaches. There are also concerns that the researchers reporting the observations may not be qualified SLPs and therefore may miss subtle speech deficits. Similarly, definitions or descriptions of the observations were not included and therefore what constitutes 'slurring of speech', for example, is unknown. Steinbok et al. (2003) also failed to report how observations were obtained from parents. Parents may have been given options, 'such as does your child talk slow', or 'does your child slur his words', or open ended questions may have been provided, such as 'describe your child's current speech'. The method of how the information was obtained will influence the validity as well as the scope of the results. According to Steinbok et al.

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(2003) “..it is reasonable to assume that if a speech abnormality was noted by the parents, it is highly probably that a speech abnormality would have been identified in a formal speech assessment” (p.182). This may be true, however the converse is not. Parents will most likely not be aware of all the aspects of speech that a trained professional would be in tune to (e.g. excess and equal stress, irregular articulatory breakdowns, prolonged phonemes and intervals, voice quality, fluency of speech) and a seemingly minor speech abnormality might not be recognized by a parent and only by a trained professional.

Another important aspect to discuss when appraising these articles is the manner in which speech samples were obtained. Huber et al. (2006) and Hudson et al. (1989) obtained connected speech samples using picture-prompted narrative speech task in which individuals were asked to tell a story or answer the question ‘what will happen next’ using a children’s picture storybook. The age of subjects in these studies ranged from 8.75 to 31.5 years and therefore the use of a children’s picture book may not have been appropriate for the older children and adults in the study. Obtaining a speech sample through these means may have resulted in shorter samples that are not as comprehensive or representative of daily speech as would have been a more open discussion with the subjects. Assessing speech in a more natural environment and through more natural means would aid in increasing the validity of the speech samples obtained. This would result in samples that were more representative of daily speech and therefore more representative of the speech difficulties that the subjects face in their daily lives, as most of the subjects probably do not make-up stories to children’s picture books on a daily basis.

It is also important to consider how speech outcomes were measured. There is a general problem reported in the literature “..in using adult criteria to classify the acquired dysarthrias of childhood, as the clinical picture in childhood is different from that in adults” (Catsman-Berrevoets et al., 1992, pg.1108). However, Huber et al. (2006) used the Dysarthria Rating Scale in order to determine the presence of ataxic dysarthria and Hudson et al. (1989) used the Frenchay Dysarthria Assessment and the method of Darley et al. (1969) to describe the speech characteristics of their subjects. Therefore, the results obtained from these studies have reduced validity and reliability.

Statistics

Inter-rater reliability was not reported by Huber et al. (2006) and therefore there is no way of knowing how many inconsistencies occurred and how often a consensus had to be reached. Inter-rater reliability scores would allow for the determination of the reliability of the results obtained. These researchers used an appropriate between-group ANOVA for each speech characteristic separately, however with only six subjects in each group it is unlikely that the study had the power to obtain a statistical difference between groups. Therefore, it is almost impossible to reject the null hypothesis and a descriptive approach might have been more appropriate for this study.

Recommendations

Based on the critical review of the available literature there is evidence to suggest that some individuals continue to have speech deficits as measured by articulation, phonology, fluency of speech, rate of speech, or the presence of dysarthria characteristics years after surgery and TCM. However, several concerns regarding the research exist including; concerns regarding recruitment of participants, small sample sizes, lack of inclusion and exclusion criteria, lack of experimental designs and control groups, and concerns regarding the use of adult criteria to classify the acquired dysarthrias of childhood. It is therefore recommended that clinicians be cautious when generalizing the findings of these studies to clients in their practice, such as when providing parents with information regarding the expected speech outcomes for their child following surgery.

It is also recommended that further research be conducted to confirm the research that has been completed and to clarify this research question. Researchers working in this area are encouraged to:

1. Use experimental study designs and include control groups.
2. Develop longitudinal studies.
3. Use objective measurements for articulation, fluency, dysarthria, and rate of speech instead of relying on subjective and descriptive approaches.
4. Include relevant and important information such as inclusion and exclusion criteria, participant histories and recruitment procedures, and complete methodological procedure for the readers to fully understand their methodology and conclusions.
5. Include more participants in their studies and use random sampling.

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6. Obtain speech samples in more natural communication settings where interactions are more spontaneous and representative of daily speech and not dependent on the task at hand (story telling).
7. Use diagnostic tools which were developed for assessing the acquired dysarthrias of childhood.
8. Research the relationship between the extent and location of cerebellar damage due to tumour resection and the extent of residual speech deficits.
9. Research the relationship between the length of the period of mutism and characteristics of the subsequent recovery and the long term speech deficits.
10. Research whether the residual deficits are so minimal as not to be perceived by the average person, and only by trained professionals through the use of diagnostic procedures.

Conclusion

The present literature review suggests that many individuals who have cerebellar tumours resected in childhood experience residual speech deficits following transient cerebellar mutism. These deficits may be of articulation, phonology, fluency, rate of speech, or dysarthria and may persist beyond two years following surgery. This information is important for clinicians to consider when providing parents with the expected long-term speech outcomes of their child following surgery and mutism. Although the research is largely descriptive in nature and contains few subjects, there were long term speech deficits in all but three participants in the previous studies. Until further experimental research can be completed, the findings from these studies can be used to show the potential for residual long-term speech deficits following transient cerebellar mutism in childhood.

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