KONGENITALE WORTBLINDHEIT ODER SPEZIFISCHE DYSLEXIE (CONGENITAL WORD-BLINDNESS)

By EDITH ZERBIN-RÜDIN, Dr. med.

In P. E. Becker (ed.) Homangenetik, Vol. V/2.
Stuttgart: Thieme, 1967.
Translated from the German by Steven G. Vandenberg, Ph.D.
University of Louisville School of Medicine.*

Dyslexia or congenital word-blindness was first described by Kerr (1897) and Morgan (1896) in children with otherwise normal intelligence. Berkhahn (1885) had not yet brought out as clearly the criterion of normal intelligence. "Congenital word-blindness" (Morgan) became the object of numerous studies. Today the expression "word-blindness" is often felt to be unsatisfactory and is replaced by the also debated term "alexia." Anglo Saxon authors at present speak mostly of "reading disability" and Scandinavian ones of "specific dyslexia." The change in the name is at the same time an indication of a change in the delineation of the disease: while in the past predominantly serious cases were considered, in which alexia persisted into adult age, as time went on milder reading problems were more often included. Most psychiatric texts discuss congenital word-blindness or alexia under mental deficiency, even though the latter is not present. Cases in which difficulties in reading and writing are due to a general lack of intelligence, or to neurotic or psychotic behavior, are not specific but secondary and do not belong here. Of course, specific dyslexia is often mistakenly regarded as mental deficiency by practitioners, and often unsatisfactorily distinguished from other types of reading problems.

Clinical Data

We are dealing with an inability of children to learn to read normally. Similar letters such as b and d are occasionally mixed up. Most of the time, however, single letters can be recognized correctly, but not combined into syllables or syllables into words.

This behavior is demonstrated by an observation of Kuromaru and Okada (1961). A typical reading difficulty occurred in a 12-year-old Japanese boy, regarding the "Kana" script, which is built up from letters and syllables in a similar way as in European scripts. In the "Kanji" script, however, which is more difficult to learn, in which every symbol stands for an entire word, the boy's reading difficulty was only minimal.

Disturbances of writing are almost always associated with the reading problem, and frequently there are also speech difficulties and left-handedness. The ability to read and write numbers is generally intact.

Characteristic is the clear distinction between the performance in reading and writing on the one hand, and on the other hand the performance in other courses, and the general intelligence of the children. Children of high ability often compensate by memorizing, and adults by practice and experience, so that in such cases the dyslexia can only be detected by testing. Neurological or brain pathological abnormalities tend to be absent (Faust 1954, Weinschenck 1962).

History

The description by Kerr and by Morgan was followed by numerous publications, of which the more important ones were summarized by Solms (1948) and by Hallgren (1950). The publications, which have appeared in great numbers in the last 20 years in the U.S. and in smaller numbers in Germany, were mostly written by psychologists or educators. Often they do not distinguish between primary and secondary reading difficulty, and don't consider other, frequently associated, problems.

Different environmental causes have been proposed, but without the demonstration of a difference from a control series which would be needed for scientific proof.

The first report of familial occurrence of congenital word-blindness is by Thomas (1905). He described two families: in one there were two brothers affected, in the other seven children and the mother. The next studies, which consisted of single pedigrees, or of single selected families, found mostly secondary cases in two or three generations. A survey may be found in Hallgren. The studies of the following authors will only be listed here: Fisher (1905, 1910), Hinshelwood (1907, 1917), Stephenson (1907), Warburg (1911), Tamm (1927), McCready (1926), Bachmann (1927), Mayer (1928), Illing (1929), Wawrik (1931), Duguid (1935), Laubenthal (1936, 1938), Rónne (1936), Mach (1937), Orton (1937), Ley (1938), Marshall and Ferguson (1939), Skydsgaard (1942), Eustis (1947), Liessens (1949), Spiel (1953), Kuromaru and Okada (1961), and Goody and Reinhold (1961).

Weinschenck (1962) reported on five males with congenital legasthenia, who were sent to the youth section of the University Psychiatric Clinic in Marburg because of serious behavior disturbances or delinquency. In only two cases had the correct diagnosis been made before. In case 5 the father was unknown, the maternal grandfather psychotic. In case 2 the father was also affected, in case 4 the father, grandfather, father's sister and two brothers were affected. The half-sibs of the proband [half-brothers and sisters of the case] from a second marriage of the mother were normal. In case 3 one sister was affected. Especially interesting is case 1: a twin sister and another sister were also affected, of the other nine siblings a female twin pair of unknown zygocity was also affected, while the remaining seven (including another twin pair) were normal.

The most thorough and voluminous study about the heredity of dyslexia we owe to Hallgren. His proband series contained 79 children

from the Stockholm Child Guidance Clinic, 43 children from a Stockholm school which gave special attention to poor readers, and 212 controls. Because 6 children were doubly ascertained, the number of probands was 116. Gripenberg (1963), in Helsinki, followed Hallgren methodologically close, but started with children of age 10 to 11 in regular schools, including classes for children with reading or hearing problems, but no children ascertained through a clinic.

The study of Malmquist (1958) is only partially comparable with the two other studies because apparently many cases of unspecific dyslexia were included. The average I.Q. of 96.6 of the 399 probands was considerably lower than Hallgren's values of 108 and 128. Malmquist tried to explain dyslexia psychologically. The suspicion can be raised, however, that the poor social and financial conditions he suggested as causes are more related to the lower I.Q. level than to the dyslexia.

Frequency

Frequency estimates run from .02% to 20%. Such a wide spread is the result of varying consideration of mild and serious cases, admixture of nonspecific, secondary dyslexia, and differing age groups and ways of collecting cases.

The German authors considered only serious cases and gave very low values. Thus Mach (1937) found only 7 cases of congenital word-blindness among 30,000 children in the Mannheim schools (0.023%), and Bachmann (1927) 10 in 51,000 children in München (0.02%). Other estimates are around 4% (f.i. Weinschenck, 1965) with the exception of Probst (1945), who estimated the frequency of alexia among school children in Basel at 0.1%. Newer authors conceive the concept quite broadly. Perhaps dyslexia does appear indeed more frequently due to the increasing use of the "whole word" method of teaching reading (Mosse 1961, 1963). Hallgren (1950) mentioned, for Stockholm, a frequency of 10% (perhaps up to 18%) of which 80% were milder cases; Gripenberg (1963) for Swedish-speaking children in Helsinki, 14.7% for boys and 7.8% for girls; Malmquist (1958) for Stockholm, 15%; Gjessing (1958) for Norway, 3.4% (only serious cases), Preston (1941) 20% for the U.S. A study conducted in 1938 by the Ross Foundation in Edinburgh found signs of specific dyslexia for approximately 10% of the pupils; however, only half of these needed special instruction.

Genetics

The incidence of secondary cases is exceptionally high. Numerous pedigrees show the disturbance in several generations. Hallgren divided his probands into 4 groups: a. both parents are dyslexic (3 probands), b. one parent is dyslexic (94 probands), c. at least one case of dyslexia among sibs, uncles, aunts or grandparents (7 probands) and d. isolated cases (12 probands). In the predominant majority (81%) there is therefore at least one dyslexic parent. Furthermore, this percentage

represents a minimum value, since adults have largely compensated for their original difficulties and often have forgotten or repressed them. A mother of one of Hallgren's probands, for instance, denied in good faith having had dyslexic difficulties — while the ensuing test brought them out clearly. For the sibs, or the children, group 2 gave the following genetic load: according to Weinberg's proband method $45.7 \pm 4.3\%$; according to Weinberg's sib method $55.2 \pm 4.4\%$; according to the method of Haldane $58.9 \pm 4.2\%$.

Gripenberg (1963) mentioned that two-thirds of her 42 cases belong to groups 1 and 2, and 95% to groups 1, 2 and 3, without analyzing her material further.

In the twin series of Norrie (1954) all 9 MZ [monozygotic, "identical"] pairs were concordant, but only 10 of the 30 DZ [dizygotic, "fraternal"] pairs. All 3 MZ pairs of Hallgren were also concordant, but only 1 of 3 DZ pairs. Single case studies of concordant MZ pairs were reported by Brander (1935); Ley and Tordeur (1936); Jenkins, Brown and Elmendorf (1937); Schiller (1937); vonHarnack (1948). A concordant boy-girl pair was described by Weinschenck (1962), and concordant pairs of unknown zygocity by Spiel (1953) and Weinschenck (1962).

Almost all authors found more males than females affected by specific as well as unspecific dyslexia. The not too numerous reports of several families or of larger pedigrees show, however, among the secondary cases a much smaller or absent excess of males. In Hallgren's series the boys outnumbered the girls by 76% (89 versus 27). Yet there were only 57% boys among the poor readers of the siblings of probands. If one puts parents and siblings together, then 92 (47%) of the males and 75 (37%) of the females are affected. The difference is still significant.

The single pedigrees alone point clearly to dominance. Hallgren also assumed transmission by an autosomal, dominant gene with almost complete manifestation. In its favor is the fact that the overwhelming majority of the probands (81%) arise from the parental combination affected x normal. His group of 12 isolated cases of dyslexia might not be uniform. In 3 cases one of the parents is suspected of dyslexia, in 8 cases environmental factors could have hindered the development of reading and writing.

Recessiveness has only rarely been proposed, for instance, by Stephenson (1907) and Tkacev (1933). Other authors assumed polygenic inheritance as in the case of intelligence. In favor of that theory would be the fact that there is a continuous transition from normal to dyslexic in which all gradations occur. The line separating normal and affected is drawn arbitrarily and cases as serious as those described by Morgan occur very infrequently. On the other hand, the situation can hardly be made to fit the Gaussian normal distribution. Furthermore, a single allelic effect seems more plausible for such an isolated defect. The

newest findings do not support the theory of Laubenthal (1938) that word-blindness is only a symptom of brain damage, which can have multiple localization and thus cause word-blindness, mental retardation and other disturbances alone or together.

Complete or partial sex linkage was roundly rejected by Hallgren on the basis of thorough calculations. The excess of boys can be explained in part by the higher manifestation and stronger expressivity in males, in part by one-sided selection of probands, especially in clinics, but also in schools.

Social conditions, such as separation of the parents, poverty, too early a start in school, being a single child, number of other children, etc., apparently are of no importance for the etiology of specific dyslexia, nor are physical illness, neurological or sensory defects. Thus environmental factors play a role in only a few cases.

Relations to Other Disturbances

Dyslexia is, as mentioned above, almost regularly accompanied by disturbances in spelling. Speech problems are more common among dyslexic children (34%) than among controls (7%), but are of a milder type. Of the brothers of probands who had reading problems (only right-handed children) 32 (41%) showed speech defects, of the normal readers 5 (13%). For the right-handed sisters, the corresponding figures were 32% and 18%. According to these results there is a direct connection between dyslexia and speech problems. The latter cannot be considered the result nor the cause of dyslexia, but both probably go back to the same basic disturbance.

Information about left-handedness in dyslexia varies; according to Hallgren this occurs more often among dyslexic children (probands and sibs) than among normals. The distinction becomes smaller after several statistical corrections, and a direct connection with dyslexia cannot be shown statistically. There were among the probands 18% left-handed boys and girls compared with 11% and 3% of the controls. Of the dyslexic sibs of the probands, 8 boys (20%) and 4 girls (13%) were left-handed, in the comparison group (control series plus normal reading sibs of the probands) the figures are 12% for the boys and 6% for the girls. The average frequency for left-handedness in adults is generally held to be 4 to 8 percent.

Investigations of the connections of dyslexia to various nervous diseases came to rather divergent conclusions, due to the absence of uniform conceptions of the nervous diseases (for literature on this see Hallgren, 1950). According to him problem children are more common (58%) among clinic probands than among their normal (25%) and poor reading (31%) sibs. Almost the same percentages were found for digestive disturbances, enuresis, headaches, sleeping problems, nailbiting and tics. At least one of these symptoms was shown by 59% of the clinic probands, by 32% of their normal and by 33% of their dyslexic siblings.

The proportions were quite similar for the school probands, except that the numbers are too small to allow generalization. Nervous disturbances are therefore more frequent among probands than among their sibs, and interestingly enough even higher than among the dyslexic sibs. There is a difference between the normal and the dyslexic sibs, but it is not statistically significant. Finally nervous disturbances occurred more often among those dyslexic children who had sought medical advice, than among those who had not.

A genetic relation to general mental retardation does not exist in Hallgren's series. Only one poor reading parent and one poor reading sibling were retarded. The idea of a close genetic relation between congenital word-blindness and mental deficiency such as was proposed by authors such as Laubenthal or Brander, resulted from the observation of single families and of especially serious cases. But especially these most serious cases of dyslexia in adults can represent a selection for low ability, because such dyslexic individuals are not able to compensate successfully for the disturbance. For the cases of Laubenthal one can also think of an accidental or selective combination of dyslexia and mental deficiency.

In summary Hallgren gave, for the siblings of the probands, the following percentages: mental deficiency 1%, nervous disturbances 21%, left-handedness 7%, speech defects 17%. These values are roughly similar to the population averages. Among the parents (dyslexic as well as normal) psychic abnormalities are no higher than for the general population.

A certain connection exists to criminality. The percentage of dyslexic cases among juvenile inmates of prisons is considerable, in American as well as in Hessen (Weinschenck, 1963) where it is about 15-20%. This is explained on the basis of the discouragement and poor adjustment that are due to depressing experiences resulting from non-recognition of their reading problem in school.

REFERENCES

Bachmann, F. Uber kongenitale Wortblindheit (Angeborene Leseschwäche). Abhandl. Neurol. Psychiat., Psychol. u. Grenzgeb. 40 (1927).

Berkhahn, O. Uber die Störungen der Schriftsprache bei Halbidioten und ihre Ahnlichkeit mit den Sprachgebrechen, Stottern und Stammeln. Arch. Psychiat. Nervenkr. 16 (1885), 78; 17 (1886), 599.

Brander, T. cited by Hallgren.

Eames, T. H. Comparison of eye conditions among 1,000 reading failures, 500 ophthalmic patients and 150 unselected children. *Amer. J. Ophthal.* 1948, *31*:713.

Duguid, K. Congenital word-blindness and reading disability. *Guy's Hosp. Rep.*, 1935, *85*:76.

Eustis, R. S. The primary etiology of the specific language disabilities. *J. Pediat.*, 1947a, *31*:448.

Eustis, R. S. Specific reading disability. *New Engl. J. Med.*, 1947b, 237:242.

Faust, C. Hirnpathologische Studie über kongenitale Schreib-Lese-Schwäche. *Nervenarzt*, 1954, *25*:137.

Fisher, J. H. Case of congenital word-blindness (Inability to learn to read). *Ophthal. Review*, 1905, 315.

Fisher, J. H. Congenital word-blindness (Inability to learn to read). *Trans. opthal. Soc. U.K.*, 1910, *30*:216.

Gjessing, R. cited by Gripenberg.

Goody, W., M. Reinhold. Congenital dyslexia and asymmetry of cerebral functions. *Brain*, 1961, *84*:231.

Gripenberg, U. Specific dyslexia in Helsinki, Finland. Proc. II. Int. Congr. Human Genet. Rom., 1961. Vol. III. S., 1936. Inst. G. Mendel., Rom., 1963.

Haldane, J. B. S. The formal genetics of man. Proc. roy. Soc. London, 1948, 135:147.

Hallgren, B. Specific dyslexia ("Congenital word-blindness"). *Acta psychiat. scand. Suppl.*, 1950, 65.

v. Harnack, G. A. Das Schulversagen als ärztliches Problem. Z. *Psychother.*, 1962, *12*:110.

Hermann, K. *Reading disability*. Munksgaard, Kopenhagen, 1959.

Hinshelwood, J. Four cases of congenital word-blindness occurring in the same family. *Brit. med. J.*, 1907, *II*:1229.

Hinshelwood, J. *Congenital word-blindness*. London, 1917; cited by Hallgren, 1950.

Illing, E. Uber kongenitale Wortblindheit (angeborene Schreib und Leseschwäche). *Mschr. Psychiat. Neurol.*, 1929, *71*:297.

Jenkins, R. L., Brown, A. W. and Elmerdorf, L. Mixed dominance and reading disability. *Ref. Zbl. ges. Neurol. Psychiat.*, 1937, *85*:519.

Kerr, J. School hygiene, in its mental, moral and physical aspects. *J. statis. Soc.*, 1897, *60*:613.

Kirchhoff, H. Erfahrungen mit Legasthenikerkiassen in Hamburg. Schulwarte, 1958, 11.

Kuromaru, S. and Okada, S. On developmental alexia in Japan. Proc. VII. int. Congr. Neurology. Rome 1961, ed. II, S. 370.

Laubenthal, F. Uber "kongenitale Wortblindheit," zugleich ein Beitrag zur Klinik sog. partieller Schwachsinnsformen und ihrer erblichen Grundlagen. Z. *ges. Neurol. Psychiat.* 1936, *156*:329.

Laubenthal, F. Uber einige Sonderformen des "angeborenen Schwachsinns" (Klinischer und erbbiologischer Beitrag zur Kenntnis der kongenitalen Wortblindheit und Worttaubheit, der Hörstörungen bei Schwachsinnigen und der xerodermischen Idiotie). Z. *ges. Neurol. u. Psychiat.*, 1938, *163*:233.

Ley, A. Sur l'alexia d'évolution familiale et héréditaire. *Ann. méd. psychol.*, 1938, *96*:145.

Ley, J. and Tordeur, G. W. Alexie et agraphie d'évolution chez des jumeaux monozygotiques. *J. belge Neurol. Psychiat.*, 1936, *36*:102.

Liessens, P. L'alexie ches les enfants arrieres. *Acta. neurol. belg.*, 1949, *49*:102.

Mach, L. Lese- und Schreibschwäche bei normalbegabten Kindern. *Z. Kinderforsch*, 1937, *46*:113.

Malmquist, E. Factors related to reading disabilities in the first grade of the elementary school. *Stockholm Studies in Educational Psychology 2*, 1958.

Marshall, W. and Ferguson, J. H. Hereditary word-blindness as a defect of selective association. *J. nerv. ment. Dis.*, 1939, *89*:164.

Mayer, K. Uber kongenitale Wortblindheit. *Mschr. Psychiat. Neurol.*, 1928, *70*:161.

McCready, E. B. Defects in the zone of language (word-blindness) and their influence in education and behaviour. *Amer. J. Psychiat.*, 1926, *6*:267.

Morgan, W. P. A case of congenital word-blindness. *Brit. med. J.*, 1896, *II*:1378.

Mosse, H. Lesestörungen in den Vereinigten Staaten. *Arztl. Mitt. (Köln)*, 1961, *58*:1626.

Mosse, H. Kindliche Lesestörungen als internationales Problem. *Arztl. Mitt. (Köln)*, 1963, *60*:428.

Myklebust, H. R. and Johnson, D. Dyslexia in children. *Except. Child.*, 1962, *29*:14.

Norrie, E. Ordblindhedens (dyslexiens) arvegang. *Laesepaedagogen*, 1954, *2*:61; cited by Stromgren, E. Trends in psychiatric genetics in Scandinavia, *in* Kallmann, F. J. Expanding goals of genetics in psychiatry. London/New York: Grune & Stratton, 1962.

Orton, S. T. Reading, writing and speech problems in children. New York: Norton Co., 1937.

Orton, S. T. Reading, writing and speech problems in children. A presentation of certain types of disorders in the development of the language faculty. *Zbl. ges. Neurol. Psychiat.,* 1939, *89*:141.

Preston, 1941, cited by Solms, 1948.

Probst, 1945, cited by Solms, 1948.

Renne, H. Congenital word-blindness in school children. *Trans. ophthal. Soc. U.K.,* 1936, *56*:311.

Ross Foundation. The W. H. Ross Foundation for the study of the prevention of blindness, Edinburgh 1938, cited by Sinclair, A. H. H. Developmental aphasia (also known as congenital word-blindness and sometimes referred to as alexia or dyslexia). *Brit. J. Ophthal.,* 1948, *32*:522.

Schiller, M. Zwillingsprobleme, dargestellt auf Grund von Untersuchungen an Stuttgarter Zwillingen. *Z. menschl. Vererb. — u. Konstit. — Lehre,* 1937, *20*:284.

Skydsgaard, H. B. *Den konstitutionelle Dyslexi.* Diss. Kopenhagen, 1942.

Solms, H. Beitrag zur Lehre von der sog. konsgenitalen Wortblindheit. *Mschr. Psychiat. Neurol.,* 1948, *115*:1.

Spiel, W. Beitrag zur kongenitalen Lese — und Schreibstörung. *Wien Z. Nervenheilk,* 1953, *7*:20.

Stephenson, S. cited by Solms.

Stephenson, S. Six cases of congenital word-blindness affecting three generations of one family. *Opthalmoscope,* 1907, *5*:482.

Tamm, A. Die angeborene Wortblindheit und verwandte Störungen bei Kindern. *Z. psychoanalyt. Pädagog.,* 1927, *1*:329.

Thomas, C. J. Congenital word-blindness and its treatment. *Opthalmoscope,* 1905, *3*:380.

Tjossem, T. D.; Hansen, T. J. and Ripley, H. S. An investigation of reading difficulty in young children. *Amer. J. Psychiat.,* 1962, *118*:1104.

Tkacev, R. cited by Solms.

Walter, K. Beitrag zum Problem der angeborenen Schreib-Leseschwäche. *Nervenarzt,* 1954, *25*:146.

Warburg, F. Uber die angeborene Wortblindheit und die Bedeutung ihrer Kenntnis für den Unterricht. *Z. Kinderforsch.,* 1911, *16*:97.

Wawrik, M. Eine experimentell-psychologische Untersuchung über zwei Fälle von Leseschwäche. *Z. Kinderforsch.,* 1931, *38*:462.

Weinschenk, C. Die erbliche Lese-Rechtschreibeschwäche und ihre sozialpsychiatrischen Auswirkungen. Schweiz. *Z. Psychol. Beih.,* 1962, *44*.

Weinschenk, C. Die erbliche Lese-Rechtschreibschwäche, ihre Diagnostik und Therapie. Bericht 5. Psychiatertag. d. Landschaftsverb. *Rheinland Bedburg-Hau,* 1963, *46*.

Weinschenk, C. Erbliche Lese-Rechtschreibe-Schwäche und Schwachsinn. *Acta paedopsychiat.,* 1964, *31*:295.

Weinschenk, C. *Die erbliche Lese- und Rechtschreibeschwäche und ihre sozialpsychiatrischen Auswirkungen.* Huber, Bern/Stuttgart, 1965.

Extracts from remarks prepared by Steven G. Vandenberg for the Research Conference on dyslexia and related disorders at Southwest Texas State College, May 15-16, 1967.

. . . My work in the last seven years has been research with presumably normal twins of high school age and with a sample of twins followed since birth. The latter are still too young to have reading problems, but some of the results with the high school age twins appear to be relevant to our topic.