ON THE DIAGNOSIS OF CENTRAL DEAFNESS IN CHILDREN

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The analysis of hearing disorders in children is a complicated business, especially so if discriminatory abilities have to be investigated. As the latter are of predominant importance in relation to eventual treatment and prognosis, special attention has to be paid to how the child interprets the signals presented to him. Speech audiometry is a useful method to investigate the power of discrimination. When speech audiometry is performed monaurally and binaurally, not only the discriminative power of one ear can be determined but the cooperation between the ears can be studied as well. Moreover, the latter method provides the examiner with a diagnostic tool to distinguish between peripheral and central deafness (Groen and Hellema 1960). This method has some limitations e.g. its applicability on young children who do not respond in any way. The child does not need to talk comprehensibly. As long as it is willing to imitate like a parrot any sound presented to him, the method is useful. Children from the age of 4 and older can be tested.

The method is based on the following assumption. Left and right ear have their first common projections in the superior olivary nucleus (Galambos). From investigations with normal hearing individuals is known that the threshold for perception is located in the central nervous system, presumably in the olive. The peripheral organs appear to function adequately even as far down as 20 db below that central threshold. (Derbyshire and Groen). Left and right ear, if identical, contribute equally if stimulated together with the same sound. They contribute proportionally if the levels are unequal (Chocholle). In speech-audiometry this would mean that the binaural articulation curve should have the same shape as the monaural one; the former should merely be shifted 3 db (or more) to the left (more sensitive) in relation of the latter (Fig. 1).

In a pathological case with an impairment of the cochlear, the central connections being intact, another form of addition should be expected. (Fig. 2).

Then the theory of compound probabilities would predict the binaural articulation curve on the basis of the co-operation of two independent probabilities.

Let the probability for perception as a function of intensity through the right

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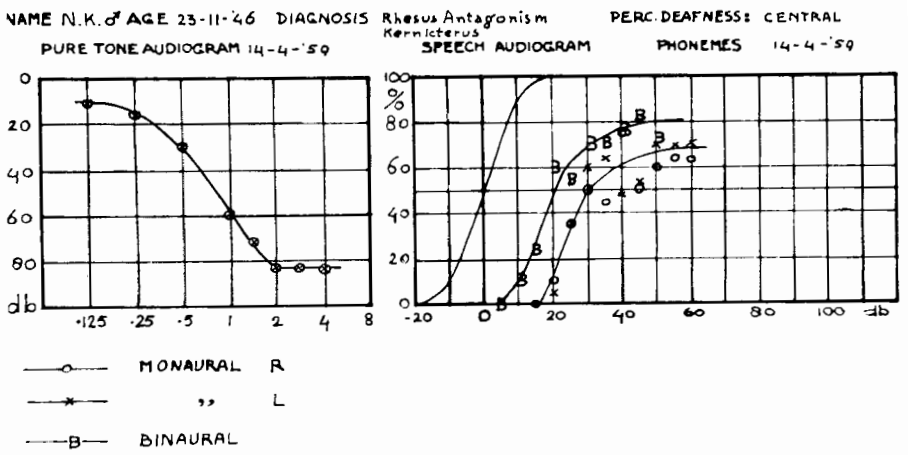

Fig. 1. Parallel shift of binaural curve relative to monaural curve. Same steepness and shape; case of central deafness.

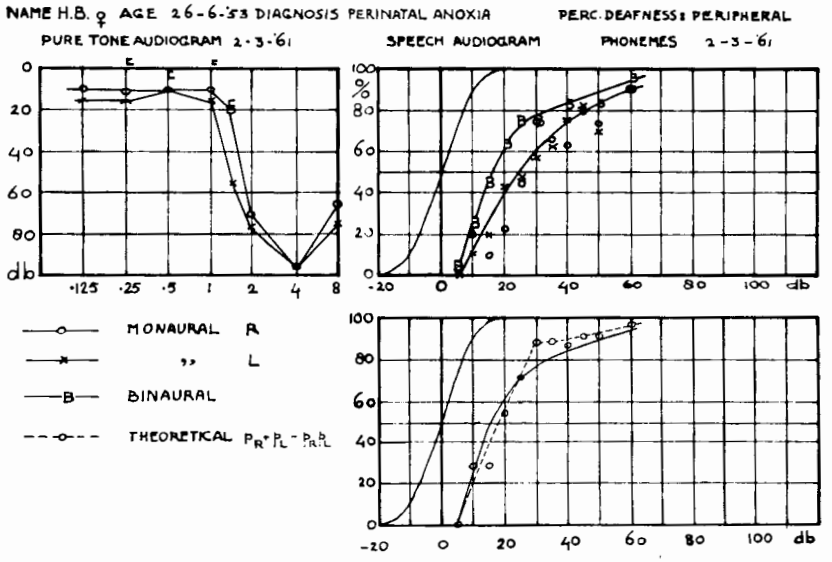

Fig. 2. Right hand top; binaural curve steeper than monaural, but starting from same point (5 db). Right hand bottom; binaural curve redrawn from top figure. Dashed line and circles pertain to theoretical binaural values, based on monaural data from top figure, according to formula $1-(1-p_R)(1-p_L)$.
Case of peripheral deafness.

ear be p_R, through the left ear p_L, (p_R and p_L between 0 and 1). Then the probabilities for non-perception either through the right ear or through the left ear would be $1-p_R$ or $1-p_L$. The compound probability for non-perception when the ears are stimulated simultaneously (diotic), would be $(1-p_R)(1-p_L)$.

The binaural probability for perception then would be $1-(1-p_R)(1-p_L)$, which expression reduces to $2p-p^2$, if $p_R = p_L$. The binaural articulation curve thus would be steeper by a factor of 2. Thus, by comparison of the shape of the binaural articulation curve with the monaural one the differential diagnosis between peripheral and central deafness can be established.

First a few words have to be added about the difference between peripheral and central. According to the assumption on binaural interaction to start no earlier than the superior olivary nucleus, every deficiency between stapes foot-plate and the olive has to be called peripheral. All other disorders located anywhere from the olive upwards have to be called central.

This method has been applied on 146 cases of purely perceptive deafness; 98 appeared to be central and 48 peripheral. The age groups of 4—10, 11—20, 21—50, 51—90 years contained respectively 50, 64, 17 and 15 cases. The majority was formed by the pupils attending the Utrecht Day School for Hard of Hearing Children. If we restrict ourselves to the children between 4 and 16 years, a group of 112 pupils, we find the following distribution of causes of deafness.

Cause of deafness	Peripheral	Central	Total
Acquired	6	14	20
Partus	5	13	18
Congenital	5	12	17
Pregnancy	6	5	11
Hereditary	1	8	9
Meningitis	5	—	5
Rhesus antag.	—	4	4
Contusio cerebri	1	2	3
Streptomycin	—	2	2
Unknown	7	16	23
Total number of cases	36	76	112

Acquired deafness: The child was born normal; measles, parotitis, flu or other toxic agents caused deafness during childhood.

Partus: Premature or retarded delivery forceps often followed by incubator, were the probable causes of deafness.

Congenital: The child has been deaf since birth, without any known reason.

Pregnancy: Rubeola, toxicosis, abnormal circumstances are probably responsible for hearing deficiences.

Hereditary: Deafness of parents or near relatives is present beyond doubt.

Meningitis: These cases are mentioned separately because they were not

treated with streptomycin. Most probably the eighth nerve has been partly destroyed, leaving the more central pathways uninvolved.

Rhesus antagonism: Of the 4 cases mentioned the existence of the antagonism has been demonstrated beyond doubt.

Contusio cerebri: In the 3 cases listed, damage to the head was caused either by a fall or by a heavy blow.

Streptomycin: The 2 cases presented here had been given streptomycin during a tubercular meningitis. Deafness may be caused by the meningitis alone, but toxic action of streptomycin may also be partly responsible.

Unknown: During childhood deafness appeared to be the reason for retarded language development. The anamnesis reveals no abnormal data about family, pregnancy, partus or early childhood.

Of the cases investigated 70% appear to belong to the group of central deafness and 30% are peripheral.

The reliability of the binaural method is demonstrated by comparing our differential diagnosis with the clinical data. In those cases where the localisation of the deficiency is known, our criteria confirm the clinical diagnosis. Hitherto no discrepancy has been encountered.

It is interesting to note that meningitis causes a peripheral deafness according to our criteria, which it should, because of the partial destruction of the eighth nerve. Rhesus antagonism appears to destroy the central pathways predominantly; streptomycin involves mainly the central nuclei, both resulting in a deafness which is, according to our criteria, mainly central.

Amongst the older patients (not listed in table) 3 cases of noise trauma, a predominantly cochlear deficiency, have all a peripheral deafness with a steeper binaural curve.

LE DIAGNOSTIC DE LA SURDITÉ CENTRALE CHEZ LES ENFANTS MALENTENDANTS

L'audiométrie vocale offre la possibilité de distinguer entre une surdité de perception centrale et celle d'origine périphérique. Les courbes d'intelligibilité monaurales et binaurales sont comparées; si la courbe binaurale possède la même forme et inclinaison initiale que les courbes monaurales, il s'agit d'un cas de surdité centrale; si la courbe binaurale est plus raide que les courbes monaurales, il s'agit d'un cas de surdité périphérique. La méthode est fondée sur la coopération binaurale qui ne commence que dans le noyau olivaire supérieur. Si la déficience est située dans la voie centrale après ce noyau les organes de l'ouïe collaborent comme chez les normaux: la courbe binaurale est déplacé parallèlement de 3 (ou plus) db en rapport avec la courbe monaurale. Si la déficience est située dans la voie, commençant après l'étrier et se terminant dans l'olive, la coopération interaurale se comporte comme le résultat des deux probabilités indépendantes, ce qui explique l'inclinaison plus raide de la courbe binaurale.

L'application en est limitée aux enfants qui veulent répondre; il n'est pas nécessaire qu'il connaissent la langue; du moment qu'ils réagissent seulement comme un perroquet la méthode est applicable. Nous avons examiné

112 enfants âgés de 4 à 20 ans (élèves de l'école des enfants malentendants à Utrecht), qui avaient tous une surdité de perception. Selon notre norme 36 des cas avaient une déficience périphérique et les autres 76 une déficience centrale. Il apparait, qu'en ces cas, où le diagnostic clinique était déterminé soit d'une surdité cochléaire ou centrale, notre diagnostic était d'accord. En 3 cas de traumatisme sonore (une déficience certainement cochléaire), nous avons trouvé une inclinaison plus raide de la courbe binaurale d'un facteur 2,0, donc appartenant à une surdité périphérique. Le Rhésus antagonisme produit des cas d'une surdité centrale. Un méningite (non traîté avec streptomycine) détruit les nerfs partiellement: en 5 cas nous avons trouvé une surdité périphérique.

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