INTRAUTERINE AUDIOMETRIC FINDINGS IN A CASE OF ANENCEPHALIA

A. PACHÌ (*), V. MONTI (*),
F. NOBILI-BENEDETTI (**),
G. L. CATASTA (**), M. R. CARDILLO (***)

(*) Department of Gyn. and Obst., University of Rome

(**) Institute of Otorhinolaryngology, University of Rome

(***) Institute of Pathology, University of Rome

SUMMARY

A study on the fetal responses to intrauterine acoustic stimuli in a case of anencephalia demonstrates a state of hyperexcitability in both the active movements and fetal heart rate, probably related to the absence of upper nervous centers.

The site of the various centers governing oto-neurological correlations in the human fetus is demonstrated.

Since the early intrauterine acoustic stimulation tests, performed by Wedenberg in 1964 ([1]), various studies have been conducted on the response of the animal ([2, 3, 4]) and human ([5, 6]) fetus to stimuli conveyed through the maternal abdomen.

The possibility of revealing functional modifications in the fetal auditory apparatus in the prenatal diagnosis of lesions and/or pathological conditions of the CNS would provide the opportunity of starting a treatment immediately at birth or possibly even earlier.

During our present research aimed at establishing, in high risk pregnancies, a correlation between extent of fetal responses and severity of the pathological condition complicating the pregnancy, we had the opportunity of studying *in utero* a case of anencephalia.

The present report deals with the fetal behaviour in response to acoustic stimuli performed according to the technique previously described ([7]).

The study of responses in malformed or neurologically impaired subjects, should offer useful neurophysiological information and provide indications on the nervous centers whether they are actively involved in the response to well defined stimuli, or in performing a particular function.

From the response of the anencephalic fetus to acoustic stimuli, it may be possible to establish the positive or negative influence upon the response of missing nervous centers.

The localization of various nerve centers implicated in the oto-neurological correlations have already been studied in experimental surgery. It would be useful to establish whether in the human anencephalic fetus the localization of these nerve centers is related to the type of malformation ([8, 9]).

CASE REPORT

D. R., aged 21, 2nd pregnancy (0-1-0-1). 32nd week. Anencephalic fetus, polyamnion. (Ultrasonographic diagnosis confirmed by amniography).

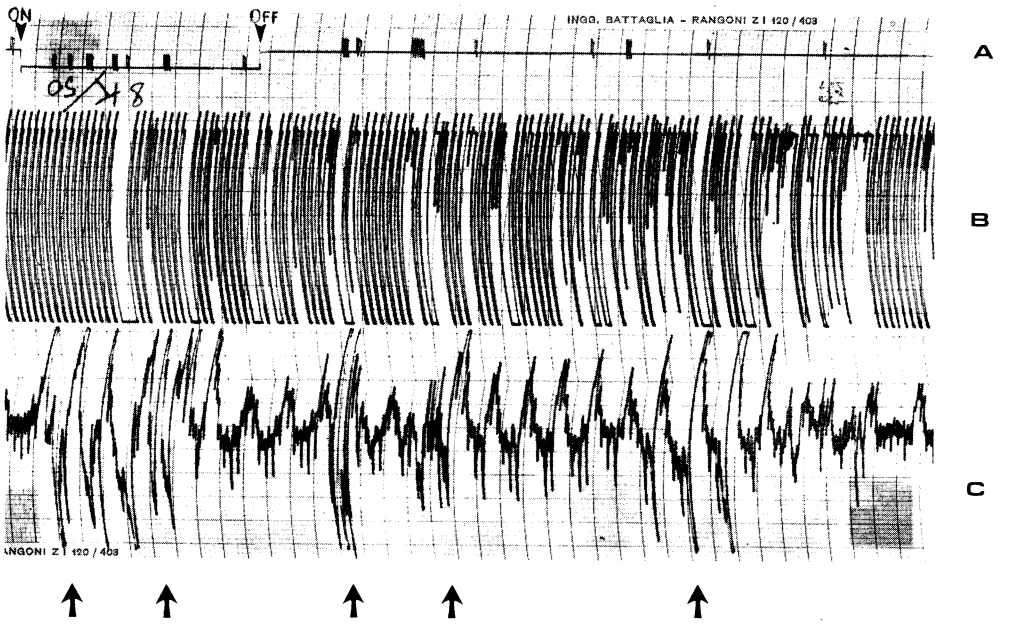

Fig. 1. — During acoustic stimulation test. Marked fetal movements (↑) were recorded by A) watch B) Fetal heart rate (FHR) and C) Rheography.

At the 34th week, spontaneous delivery of a female fetus, weighting 1600 g, which died 5 minutes after birth. Placenta 450 g.

FETAL AUTOPSY FINDINGS

Autopsy performed at the Institute of Pathology, University of Rome (file No. 56569), revealed the following data:

Head: Cranial vault, completely absent. Squama frontalis, parietalis, and occipitalis, rudimentary. Occipital hole, open posteriorly. Base of skull, reduced in size. Pyramids of the temporal bone, short and located transversally.

Cranial cavity: Telencephalon, diencephalon, mesencephalon and rostral portion of rhombencephalon, absent. Base of skull covered with a thin spongy, vascular membrane. Rudimentary cranial nerves, present. Medulla oblongata and medulla spinalis, thin and poorly developed.

Heart: Normal shape and volume.

HISTOLOGICAL PATTERN

1) Cerebrovoscular area: composed of connective tissue and collagen scattered with numerous bloody lacunae, glial agglomerates, choroidal plexuses and hemorrhagic areas. Rudimentary cranial nerves are present.

2) Medulla: white matter reduced compared with grey matter, particularly the lateral bundles. Central canal irregular in shape. Grey matter characterized by a reduction in nerve cells and by an increase in glia. Dura mater thickned and pia mater amply vascularized.

AUDIOMETRIC FINDINGS

Acoustic stimulation tests were performed at the 33rd week of pregnancy. The technique employed has been described elsewhere (7).

Once regularization of the fetal heart rhythm was established, acoustic stimulation with pure tones was performed. The first responses were obtained with stimuli of 50 dB which induced marked variations in fetal heart rhythm (mainly tachycardia), and active fetal movements, detected both by the rheograph and simultaneously by the mother (fig. 1).

While the cardiac response was immediate, the active movements started, only after a few seconds of stimulation and continued gradually increasing even during the pauses between stimuli.

With acoustic stimuli of 50 dB intensity, habituation phenomena could be observed only for active movements and only after repeated stimuli. At very high intensity (90 and 110 dB), responses were paradoxically less accentuated and limited to the per-stimulatory period.

Repeated stimuli, with sounds of high intensity, instead of attenuating, actually accentuated the response, particularly the fetal movements.

DISCUSSION

The responses obtained in the anencephalic fetus still not differing much from those recorded in healthy fetuses of the same gestational age (minimum threshold response at 50 dB vs 30 dB normal fetus) were particularly accentuated, and showed no habituation phenomena to prolonged high-intensity stimuli.

Habituation to low-intensity stimuli may be regarded, as a normal acoustic protection measure merely at the organ level or, more probably, a "fatigue" phenomenon of Corti's organ.

Studies on the neonatal period were not possible in the present case of anencephalia on account of the very brief survival time. However the peculiar anatomical and neurological lesion in this particular case has allowed us to make some considerations on the specific responses of the anencephalic fetus to acoustic stimuli.

Already in 1947, Monnier and Willi ([10]) in studies on anencephaly newborns that lived only for a few hours, noticed that, in absence of telencephalon, mesencephalon and rostral portion of the rhomboencephalon, the medulla oblongata and medulla spinalis represent the CNS.

Functions in the malformed newborns of this type are limited to those of a primary nature and the survival period varies from case to case depending upon the type of anatomical malformations ([11]).

The formation of the cerebrovascular area is possibly due to the absence of the skull and skin, leading to degeneration of the neural tissue, which is replaced by a spongy and vascular mass ([12]).

The state of hyperexcitability observed at birth is probably similar to that detected during and immediately after the intrauterine audiometric tests.

This state is probably due to functional deficiency of the higher nervous centers which in the normal fetus function regularly after the 28th-30th week of pregnancy.

It is not possible on the basis of a single observation to offer an explanation for the hyperactive movements of limbs, and in particularly the variations in fetal heart rate.

These might be interpreted, as pointed out by Sokolov ([13, 14]), as "self-protection movements".

In fact Sokolov observed that in the newborn, the cardioacoustic reflex is added to the orientative reflex, where by, in some cases "reinforced responses" can be obtained that are usually manifest as tachycardia.

Finally, these reactions, already observed in decerebrated experimental animals, may be simple automatic spinal movements ([15, 16]).

BIBLIOGRAPHY

1) Wedenberg E., Johansson B., Westin B.: *Acta Oto-Larying, 57*, 188, 1964.
2) Walker D., Grimwade J., Wood C.: *Am. J. Obst. Gyn., 1*, 91, 1971.
3) Bench R. J., Vass A.: *Lancet, 10*, 91, 1970.
4) Henshall W. R.: *Am. J. Obst. Gyn., 15*, 576, 1972.
5) Murphy K. P., Smyth C. N.: *Lancet, 1*, 972, 1968.
6) Scibetta J., Mirtimer R., Charles J., Lawrence C.: *Am. J. Obst. Gyn., 1*, 82, 1971.
7) Pachì A., Monti V., Nobili-Benedetti F., Catasta G. L.: *Pat. Clin. Ost. Gin., 5*, 237, 1976.
8) Wilder J.: *J. Nerv. Men. Dis., 125*, 73, 1957.
9) Reese W.. Dykman R.: *Physiol. Revies, 40*, 250, 1960.
10) Monnier M., Willi H.: *Ann. Paed., 196*, 289, 1947.
11) Kennet K., Nakano S.: *Child Neurol., 15*, 383, 1973.
12) Dekaban A. S.: *J. Neuropath. Exp. Neurol., 22*, 533, 1963.
13) Sokolov E. H.: *Neuronal models and the orienting reflex. M.A.B.* Edited by Brazier. The central nervous system and behaviour. New York, 187, 1960.
14) Sokolov E. H.: *Ann. Rev. Physiol., 25*, 545, 1963.
15) Dykman R. A., Gantt W.: *Am. J. Physiol., 185*, 495, 1956.
16) Wilson R. S.: *J. Compar. Physiol. Psychol., 58*, 290, 1964.